Treatment of Heart Disease

A CLINICAL PHYSIOLOGIC APPROACH

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W B SAUNDERS COMPANY

Philadelphia London 1956

Made in the United States of America at the Press of
W B Saunders Company, Philadelphia

LIBRARY OF CONGRESS CATALOG CARD NUMBER 56-8027

PREFACE

There has been no dearth of books on cardiology Numerous texts have been written on electrocardiography, cardiac roentgenography, physiology, and even treatment of heart disease The question must naturally arise, Why another?

The study of cardiac disease has long passed the descriptive phase It is now pos sible to determine the mechanisms of symptoms and the physiologic basis of the efficacy of various forms of treatment After long years of cardiac practice and of teaching cardiology to both graduate and undergraduate students it seems to us that only by understanding these mechanisms can the practice of cardiology reach the level of effectiveness made possible by advances resulting from theoretical and experimental work

However, this is not a book on cardiac physiology, nor is it a compilation of signs symptoms and therapy of heart disease It is, rather, a presentation of therapy in heart disease based upon sound physiologic principles

It is our intention to reach the general physician who imbued with the physiologic point of view may more readily understand the symptoms and clinical course of his patients

This volume is divided into seven parts and an appendix The first part consists of cardiac physiology following sections deal with degenerative diseases, inflammation of the heart or the results of infection con genital anomalies effects of pregnancy and surgery on the heart effects of metabolic disturbances and vitamin deficiencies on heart disease, and finally, the adjustment to emotional disturbances and the problem of rehabilitation

Current means of managing the arrhyth mias, the uses of digitalis, quinidine, mercurials, and diets are discussed in detail The present-day management of con gestive failure, both in its acute and in its chronic form and the complicating electrolyte imbalance are explored The am bulatory treatment of heart failure which makes it possible for patients with this disease to remain integrated in society is thoroughly discussed In the section on degenerative diseases a chapter is devoted to a discussion of hypertension including the recently developed antihypertensive blocking agents Another chapter is de voted to the ever present and increasing problem of arteriosclerotic heart disease with a consideration of its medical and surgical management

The role of the antibiotics and the steroids in the prevention and treatment of rheumatic fever and other inflammatory heart diseases is discussed The pitfalls of treatment failures and adverse reactions to drugs are evaluated In this section a detailed discussion of criteria for surgical intervention in rheumatic heart disease is presented

The treatment of congenital heart dis ease has recently undergone thorough revision The techniques of localization of a lesion by catheterization angiocardiog raphy and aortography have been com

bined with the imaginative and ingenious advances of surgery to restore many of these hearts to good functional capacity and some even to normal The combination of these techniques has rescued many patients from the half life of the congenital cardiac This long hoped for advance has also been thoroughly discussed

The effects of anesthetic agents on cardiac rhythm and myocardial function are presented The tendency of some of these agents to produce anoxia and the resulting ill effects, are considered individually The choice of anesthetic agents in relation to heart disease is discussed Measures for the prevention and management of shock are considered Consideration is given to the dangers of surgery in the cardiac patient and a clinical approach to the management of the patient who requires operation is presented

Cardiac disease in pregnancy is divided into several categories The management of each group is discussed in relation to the physiologic changes caused by pregnancy

The physiologic background of disturbances of thyroid function on the heart is detailed together with current medical and surgical treatment of these disorders

In keeping with present trends the role of emotional problems in the production of cardiac symptoms is presented in the chapter on 'Psychosomatic Aspects of Heart Disease Means of preventing and treating such problems are explored In the same section, in a chapter devoted to "Living with a Sick Heart, the interrelationships of the patient and his family are discussed The text closes with a chapter on 'Rehabilitation which deals with the maintenance of the patient s place in his family and in the community

The illustrations graphically present many of the points which are stressed in the text The diets, menus and recipes were obtained from hospitals and patients and should offer sufficient variety and ease of preparation to make eating a pleasure rather than a chore for the patient While not intended to vie with the excellent diet books now available, these suggestions should greatly aid the physician who must guide the patient's diet

In the preparation of this volume many colleagues and friends gave inestimable help It is a pleasure to thank Dr Louis Leiter, Chief of the Medical Division at the Montefiore Hospital, who read the entire manuscript and gave much helpful guidance We also gratefully acknowledge our indebtedness to Dr Arthur Fishberg for critical evaluation and encouragement, to Dr Jacob Grossman, who helped us tremendously with the chapter on physiology and to Dr R E Weston who furnished valuable graphic material Doctors A E Bloomberg, A S Buchberg J Clahr, C D Enselberg, S Gartner, E S Hurwitt, Belle Jacobson, Edith Kepes, M. Moser, Eli H Rubin, Ira Rubin, J B Schwedel, and D Young also helped greatly with criticism and discussion We are indebted to Drs Enselberg Ira Rubin, and Young for the use of some of the electrocardiograms, and to Dr Robert F Rushmer for permission to use illustrations from his book, 'Cardiac Diagnosis A Physiologic Approach

We wish to express thanks to Miss Lynn Perry and Miss Roslyn Hacker, who reviewed the chapter on diets, and to Mrs Florence Hess, Miss Ann Jezer, Daniel A Jezer and Mrs Beatrice Gross Atkin who gave invaluable help in the preparation of the manuscript We also express our thanks to W B Saunders Co, whose staff made much of the task lighter than it might otherwise have been

Finally we acknowledge most happily our great indebtedness to Montefiore Hospital, which provided the incentives and opportunities that made this book possible

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Part I

THE BASIC MECHANISMS OF CARDIAC SYMPTOMS AND THEIR MANAGEMENT

Chapter 1

PATHOLOGIC PHYSIOLOGY OF MYOCARDIAL INSUFFICIENCY

In this chapter we are concerned with contractility, the most important physiologic function of the heart Contractility, when adequate for peripheral needs, is synonymous with cardiac compensation and, when impaired, with myocardial insufficiency The other cardiac properties—rhythmicity, irritability, tonicity and automaticity—are beyond the scope of this chapter

It is the propulsive force deriving from contractility that supplies blood to all the tissues of the body As in any other pump mechanism, the efficiency of the myocardium is expressed as the ratio of energy expended in performing useful work to the total energy released by cardiac contraction

The energy for the contraction of the heart muscle is obtained by the breakdown of various chemical compounds, and has been calculated to be 1 kg m for each cubic centimeter of oxygen consumed The mammalian heart uses 1 2 to 1 8 cc of oxygen per gram per hour at rest and 2 to 6 cc with exercise Variations in oxygen usage are great The ratio of physical work accomplished to oxygen used represents the mechanical efficiency of the heart [See references 24 and 52]

The energy derived from contraction of the heart is expended in moving a column of blood and in this movement overcomes resistance which opposes the flow of blood through the blood vessels The energy expended in systole must be restored in diastole Normally up to 95 per cent of this energy is dissipated as heat The remaining 5 per cent is used to perform mechanical work

Contraction is in part controlled by the ability of heart muscle to form actomyosin for contraction and splitting into myosin-ATP and actin for relaxation In the conversion of actomyosin, work is accomplished The dephosphorylation of adenosine triphosphate (ATP) to adenosine diphosphate (ADP) probably is associated with relaxation and restoration of energy before the next contraction For this process to continue efficiently phosphocreatine must be present to give up a phosphate bond for the reformation of actomyosin ATP This exchange requires the presence of potassium Sodium has an effect opposite to that of potassium inhibiting the transfer of the phosphate bond and interfering with muscle contraction It should be emphasized that these observations pertain only to animal experiments [See references 30, 53 and 108]

Oxygen and nutrients are delivered to the heart by way of the coronary arteries The venous blood of the coronary circulation contains only about 4 to 7 volumes per cent of oxygen or 20 to 30 per cent of the arterial concentration The oxygen tension of the myocardium is, therefore, normally low During exercise when extraction of oxygen is even greater than normal the oxygen tension of the heart can be maintained only by

an increase in blood flow through the coronary arteries The blood flow through the coronary arteries depends upon a number of factors Chief among these are

1 The pressure gradient between the capillaries and the right atrium in a greatly enlarged right atrium under increased end-diastolic pressure due to failure, there may be an impediment to inflow due to this tension [See reference 6]

2 Vasomotor regulation stimulation of the sympathetic system causes a decrease in systolic coronary flow and an increase in diastolic coronary flow, an increase in extraction of oxygen and an increase in work of the left ventricle The vagus maintains the tone of the coronary arteries and stimulation of the vagus causes coronary constriction [See references 37 and 38]

3 Pressure on the intramyocardial arteries in systole greatly curtails but does not prevent coronary flow The coronary flow in diastole is more than double that in systole [See reference 39]

4 Local metabolites may influence coronary flow by causing vasodilatation Anoxia is the chief cause of coronary vasodilatation The *pH* and CO_2 tension are not major factors in determining coronary flow [See reference 88]

Oxygenation of Tissues

The quantity of oxygen taken up by the tissues depends upon two factors (1) the flow to the tissues and (2) the fraction of oxygen removed, or, in other words the mean arteriovenous oxygen difference The oxygen delivered at rest is about 250 cc per minute since about 45 to 50 cc per liter are removed from an average cardiac output of 5 liters Smaller fractions are removed when the cardiac output is large Even when the cardiac output is reduced, the blood supply to vital centers, though not necessarily to the brain, is maintained at the expense of such organs as the kidneys, which tolerate well a reduction in blood supply The cerebral blood flow in heart failure has been found to be reduced by as much as 39 per cent at the same time that the mean arterio venous oxygen difference was 41 per cent above normal The mean cerebral oxygen consumption was also found to be reduced [See references 13 and 89]

CARDIAC OUTPUT

Determination of Cardiac Output

THE FICK PRINCIPLE No direct method is available for determining the output of the heart Numerous indirect methods have, therefore, been devised Of the various techniques tried, the one utilizing the Fick principle is the most reliable According to the Fick principle, cardiac output can be calculated by dividing the oxygen consumption by the mean arteriovenous oxygen difference If the quantity of oxygen delivered by a specific amount of blood to the tissues or removed from the lungs and the total quantity of oxygen used up in a specific time are known, the volume of blood that has carried this quantity of oxygen can be determined

The procedure is as follows (1) The oxygen consumption per minute is measured by the Tissot or Douglas bag (2) The arterial and venous oxygen contents are determined and the oxygen content of the mixed venous sample is subtracted from the oxygen content of the arterial blood The resulting difference is the arterio venous oxygen difference, ordinarily expressed in cubic centimeters of oxygen per 100 cc of blood (3) The cardiac output is determined by dividing the total quantity of oxygen consumed in 1 minute by the arteriovenous oxygen difference The quotient is the volume of blood necessary to transport the amount of oxygen utilized per minute For example, if the oxygen consumption is 260 cc per minute and the oxygen difference between arterial and mixed venous blood is 5 cc per 100 cc, the cardiac output would be $\frac{260}{5} \times 100 = 5200$ cc, or 5 2 liters per minute

Recently catheterization of the coronary

sinus has made it possible to remove samples of blood from the myocardium and to determine oxygen consumption and the efficiency of the heart muscle under experimental conditions [See reference 7]

According to Cournand the validity of the Fick principle cannot be challenged if it is admitted that (1) small inequalities in both ventricles are cancelled over a succession of cardiac beats and respiratory cycles, (2) the measured output is actually the effective ventricular output and does not include blood regurgitated through the A V valves and (3) the oxygen displaced from the inspired air is not utilized in the lungs for any purpose other than transport in the circulating blood [See reference 43]

Though there are inherent sources of error in this method it is of practical value when it can be done under constant or steady state conditions [See reference 114]

THE PULSE PRESSURE METHOD The pulse pressure multiplied by the heart rate has been used in determining cardiac output This method has an error of the magnitude of 25 per cent or more The sources of error inherent in this method are so great that it has little clinical value [See references 85 and 88]

BALLISTOCARDIOGRAPHY Ballistocardiography has also been used to determine cardiac output using the size of the I and J waves as an index of this function The curve is determined by the blood velocity acceleration or displacement in the large vessels, depending upon the instrument used The volume of the blood ejected is not reflected on the ballistocardiogram This method for determining cardiac output is not clinically reliable [See references 42 and 101]

THE DYE DILUTION METHOD Cardiac output is determined by the dye or isotope dilution method by observing the magnitude and timing of the rise and fall of the peripheral concentration of a test substance during its first circulation following injection into a peripheral vein The formula for the method is $F = A/Ct$, where F is flow, A is the quantity of the dye injected C is the average concentration of dye in the sample and t is the duration of sample withdrawal

This method may yield fairly reproducible results, but they do not correspond in some studies with the output derived from the application of the Fick principle [See references, 17, 60, and 76]

Normal Cardiac Output

The data as determined in various laboratories by the Fick principle, show the average normal adult cardiac output to be about 5 4 liters per minute This can be restated in terms of output per minute per square meter of body surface called the cardiac index The average normal cardiac index is about 3 1 liters

Numerical values for cardiac output are better appreciated in the light of the blood flow requirements of various important parts of the body Careful measurements indicate that the average blood flow at rest through the various regions is as follows

Heart	5400 cc per minute
Brain	15%
Kidneys	18%
Liver	18%
Extremities	30%
Remaining organs	19%

Flow to the lungs is slightly higher than the flow to the peripheral system on account of the bronchial circulation

Both in health and disease, cardiac output may be greatly augmented to meet increased demands on the heart under various conditions There is a very sharp rise after exercise and outputs in excess of 30 liters per minute have been recorded The rise in output due to anxiety may also be marked reaching as much as 15 liters per minute Change from the recumbent to the upright position causes fall in the cardiac output by as much as one third [See references 51 and 86]

stance the extrovert is predisposed to an affective or manic depressive reaction introvert may develop schizophrenia

The complaint which the patient makes to his physician may be revealing defect in temperament

Psychopathic Personality If an individual possesses mental inadequacies deviations which are neither psychotic nor feeble minded and if these aberrat render him unable to participate in satisfactory manner in social and cultural r tions with those about him the condition is known as psychopathic personality

Psychiatrists define various types of psychopathic personalities for instance stable excitable impulsive egocentric liars swindlers antisocial and quarrelsc (Kraepelin) Here however mention will be made of pathologic liars and swindl antisocial personalities sexual psychopaths and persons with situational psycho:

Pathologic liars and *swindlers* are of agreeable manners and optimistic Tl select as their dupes the credulous with whom they make social or business conta They distort facts about themselves concerning their own attainments and kno edge in order to profit at the expense and humiliation of their victims The credulc individuals for example maids and matrons alike often are blackmailed for sex fenses Money and sums from other sources may be obtained by the swindlers su posedly for investment purposes The pathologic liars and swindlers are unsta and are incapable of accepting responsibility or are unwilling to do so

The individual who has an *antisocial personality* is affectively cold and has acceptable constructive expression of the striving functions He or she rarely fe remorse at offenses against person property society or law Many take pleasure a feel pride in their ability to offend moral religious and legal standards

Sexual psychopathy is manifested in some by an abnormal sexual goal as homosexuality and in others by the gratification of the impulse by abnormal metho as in sadism masochism and exhibitionism

Many of the so called *situational psychoses* represent schizophrenic affective paranoid reactions which occur under circumstances involving great emotional di tress and they thus represent attempts to escape from the hard and uncompromisin reality of some specific difficult situation Frequently situational psychoses are the nature of confused states or paranoid episodes or of attacks of irritability ex citement or depression

Patients afflicted with the difficulties described may complain of them or the may try to conceal their difficulties and the complaint may be made by others Like wise in any of the psychoses the complaint may be expressed by the patient or mad by the family or friends of the patient

Complaints of the abnormalities just described are less likely to be made by th patients than by relatives by those duped or by officers of the law

Character Disturbances and the Psychoses The psychic functions in ma have evolved from development of the simple instinctive tendencies In each instinc tive tendency there are the constituents of knowing (cognitive) feeling (affective) and striving (conative) Feeling and striving are considered as character as con trasted with cognition and intellect The intellect seems to be an instrument which has been evolved in order better to satisfy man s desires by activities through know ing In order that a human being may know how to adapt and respond to the ever changing forces of physical and social environment there must be ever present adequate cognitive affective and conative functioning of the mind

Complaints of psychic origin often represent faulty technics of adjustment neu rotic compromises with reality and the results of various protective and safety devices employed by the patient in order to serve the ends and aims of the person ality In order to understand the complaints of a mental disorder one must have an understanding of the forces that have gone into their formation as a means to likely ascertaining what the patient is reacting to and expressing in his com

In certain instances the services of the psychiatrist may be required for a full elaboration

Knowing (Cognitive) Defects Complaints originating as a result of disturbed functions of cognition and intellect are referable to (1) the sense organs (2) the neurons of peripheral nerves which conduct impressions through the central nervous system to the higher centers of integration and (3) the integration and combination of these new impressions with old ones to form perceptions and judgments that are in agreement with objective reality How far a given human being the patient is healthy or ill and the reason therefore depend on the degree of disturbance of the psychic and the somatic functions It is desired to determine how far the patient is capable or incapable of making adequate responses to the psychic and physical influences of the environment in which he lives

THE SENSE ORGANS The nerve beginnings in the organs of sense namely the eyes ears nose taste buds touch spots in the skin and the nerve beginnings in the muscles bones joints and viscera are essential to the functions of knowing

The various end organs or receptors are so constructed as to analyze the environmental energy and select certain kinds which are transformed in the receptor and give rise to a nervous impulse On arrival in the appropriate preordained area in the brain this impulse produces a visual auditory or other sensory image the interpretation and meaning of which will depend on the previous experiences of the interpreter The elements which are likely to lead to misinterpretations of these images with resulting illusions are deeply intense affective states or strongly urgent drives and impulses A misinterpretation of normal stimuli arriving at the higher centers of integration is an *illusion* The nature of illusions is likely to be determined by the prevailing trend of the patient s preoccupations In confused toxic states visual perceptions are more likely to be misinterpreted than perceptions involving other senses because the sensory stimuli and impressions are not sharply defined thus the common occurrence of visual illusions in alcoholic and other intoxications

A more serious falsification of perceptions is the formation of image symbols when there is no discoverable impulse created by the stimulation of a receptor A mental image projected outward as a perception in the absence of an external stimulus is an *hallucination* An hallucination lacks a basis of reality It is created by the patient s own mind but it constitutes an actual part of the mental life of one thus affected

The content of perceptions in hallucinations is so intimately subjective that the patient cannot ignore them They absorb the attention completely and reality is made to harmonize with them The nature of the material hallucinated is influenced by the psychologic experiences of the patient

During the early and acute stages of a toxic psychosis hallucinations are more marked and may be of no great importance in these disorders The hallucination which occurs with a clear mind is of serious diagnostic import Hallucinations are frequent when attention slackens and there is time for daydreaming

In hallucinations the images are cast in that sense best fitted to symbolize them Hallucinations of *hearing* are most frequent Feelings of guilt for example may best be expressed in spoken language and so the patient hears accusing voices The voices arrange the words in sentences and give commands addressed personally to the patient The commands are often convincing and compelling and may therefore coerce the patient to overt acts

In the deliria of acute infectious diseases or of toxic psychoses hallucinations of *sight* occur most frequently The classic form of mental disease accompanied by visual images is delirium tremens The images are almost always of a terrifying nature

Olfactory hallucinations most commonly represent feelings of guilt and often occur in schizophrenic states

Hallucinations of *taste* are uncommon and when present they often are associ ed with hallucinations of smell

Tactile hallucinations are common in toxic alcoholism and cocaine addiction

Sexual hallucinations are almost always limited to schizophrenia The hallucina ons pertain to size shape or the absence of the external genitalia

All of these abnormalities *related to the sense organs are likely to be perceptible* i the patient s complaint as he tries to express it to the physician

THOUGHT The joining of ideas and the formation of new ones constitute the inction known as thinking Thought processes may be disturbed in production rogression and content

In mental health the stimuli for thought come from environmental reality and hus thought is rational or realistic In mental disease the production of thought omes from the inner mental life and is not corrected by reality For instance schizo *phrenic thinking finds its origin in complexes* wishes *drives* and other motivations not recognized by the patient and operates for the purpose of providing substitute forms of satisfaction Thought wanders without correction by realities

Disorders in Progression of Thought The rate and manner of progression of thought if revealed at all are disclosed by the stream of talk Of course it is expected that during normal small talk for the purpose of entertainment there may be a lack of logical and coherent sequence of related ideas Beyond this small talk for enter tainment purposes there may be disturbances of the stream of thought in which the thinking processes appear to run too quickly and in which at least some ideas are not completed This condition is known as *flight of ideas* In flight of ideas words of similar sound call up the new ideas This new association of ideas is known as *clang association* and may lead to a pun or a senseless rhyme

The opposite to a flight of ideas is *retardation* of the thought processes In re tardation the initiation and movement of thought are slow The patient s thoughts come slowly and are expressed by a slow speech and in a low tone Retardation in minor degrees may be normal when fully developed however it is observed chiefly in the depressive phases of the affective psychoses but also may be present in schizophrenia

A disorganization of syntactic structure of thought with a lapse into disjointed phrases or even into parts of sentences is known as *incoherence* Incoherence occurs particularly in schizophrenia a disease in which the thinking is characteristically dominated by complexes

When both expression and progression of thought suddenly but temporarily cease the process is known as thought obstruction or *blocking* This often occurs in petit mal epilepsy

When there is a tendency for the associative evolution of ideas which are deter mined more by affective factors than by logical reasoning the strongest feeling tones dominate An idea which comes to have an extreme feeling tone connected with it is spoken of as an *overdetermined* or *overvalued* idea An overvalued idea is an im portant determinant of behavior Individuals possessing degrees of overdetermined ideas are frequently met, particularly in scientific circles and religious gatherings

A *delusion* is a false conception of a particular life situation The trend of a de lusion is determined by the prepsychotic personality problems of the patient The sources of these problems may often be found in thwarted trends and drives frus trated hopes feelings of inferiority inadequacies impelling desires feelings of guilt and other affective preoccupations

The psychologic purposes and needs for the satisfaction of which delusions are created fall into groups of well defined mental mechanisms For instance delusions of grandeur arise from feelings of inadequacy insecurity or inferiority any conscious recognition of which is prevented by the exaggerated affective components of the

delusion Delusions of self accusation may arise from the repression of unacceptable trends and desires

Unworthy desires or troublesome and disowned aspects of the personality when projected as hostility from the environment are termed a *delusion of persecution* Delusions of persecution permit a shifting of responsibility Those in whom delusions of persecution develop have always been too critical resentful suspicious unhappy lonely brooding and insecure from a lack of friends with whom they could share confidences

When an individual interprets the remarks or actions of other persons although in no way referring to the observer as being significantly related to himself and particularly if to him these remarks and actions express accusation or depreciation the condition is termed *ideas of reference* In paranoid states ideas of reference represent a projection of self-criticism onto the external world In depressed states feelings of guilt often stimulate ideas of reference and may contribute to loosely organized paranoid delusions

When repression weakens memories or ideas which would cause emotional pain if admitted to consciousness may give rise to depression A *depressive delusion* is the rationalization of a sense of depression resulting from unconscious hostile tendencies felt toward innocent persons

If an individual s attention is abnormally concentrated on his own body so that he is depressed and his thoughts are obsessively preoccupied with some bodily organ which is thought to be incurably diseased the condition is known as *hypochondria* Hypochondria often occurs in those who have shown a tendency to evade the responsibilities of life through illness Hypochondriasis occasionally is symbolic as in schizophrenia and as such it indicates a serious disorder

An *obsession* is the result of thoughts which persistently thrust themselves into consciousness against the conscious desire of the patient Obsessive thoughts may lead to compulsive acts In a compulsive act the patient feels impelled to perform it The act is often elaborate and repetitive so that it seems almost ritualistic

Phobias are like the obsessive idea The individual is filled with morbid anxiety and distress which constantly thrust upon him against his own will Among the common phobias are fears of dirt of bacteria of cancer of crowds or of closed spaces

Relationship to the Complaint If the physician has the confidence of his patient the foregoing difficulties in relation to thought if they have been experienced are likely to be mentioned in the patient s complaint

MEMORY The memory may be phenomenally good *hypermnesia* or there may be a loss of memory or *amnesia* and in many instances there is falsification of memory or *paramnesia*

Hypermnesia Normally the impressions which arise from events which arouse the emotions are registered with more than the usual intensity with the result that there is always a more vivid recollection of details of these events

Amnesia In instances of physiologic disturbances of neurons from toxins trauma or degenerative changes there may be organic amnesia Organic loss of memory is due mostly to an impairment of retention though registration is somewhat affected In the patient who has psychogenic amnesia the absence of memory is an active defensive process The patient always is a hysteric who simply refuses to remember

Paramnesia Paramnesia may be of different forms 1 In *confabulation* the gaps in memory are packed full of fabrications which are without any basis of fact Confabulation is observed in Korsakoff s syndrome but occasionally also in senile psychosis 2 *Falsifications* or *illusions* of memory are created in response to affective needs They are frequently observed in the reporting of past events In paranoid psychoses this tendency is highly developed 3 In the phenomenon of *jamais vu* there is a false feeling of unfamiliarity with situations that have actually been expe

ed These phenomena may occur in schizophrenia psychoneuroses epilepsy and in states of fatigue or intoxication

Dementia Dementia is a permanent, irreversible loss of intellectual capacity It curs most frequently in cerebral atherosclerosis and neurosyphilis

Disturbances of Consciousness Disturbances of consciousness are often asso iated with the acute stages of toxic infectious or traumatic conditions and some l diseases A patient in a state of *confusion* is bewildered perplexed and dis iented The face presents a distressed and puzzled expression

In *psychogenic stupor* there is no suspension of consciousness but the patient oes not move The thoughts are dereistic In toxic organic stupor conscious thought ocesses are suspended

Delirium is usually associated with fever or toxic states There is a clouding of onsciousness with bewilderment restlessness confusion and disorientation and llusions and hallucinations may occur

Dream or twilight state is an affective disturbance of consciousness in which the ient does not recognize familiar surroundings In response to visual and auditory allucinations the patient may run away from home or commit acts of violence When al consciousness is regained the patient may report that he felt as if he were ng and will have little or no recollection of events that occurred during the stence of the twilight state Such dream states occur for the most part in hysteria in epilepsy They may end in delirium as described or they may pass into dis ociated states such as fugues and double personalities

Disorders of Apperception Disorders of apperception occur in mental deficiency in psychogenic diseases involving intense preoccupation and in toxic and organic In disturbances of apperception the patient has difficulty in understanding estions and unaccustomed situations and experiences

Disorders of Orientation If a person does not know his position in reference to ime does not appreciate the situation both as to space and circumstances and does not understand his relationship to other individuals he is said to be disoriented An dividual may be disoriented in any one two or all of these spheres that is the ime place or person Disorientation may occur in any mental disease in which there extensive impairment of the memory

Relationship to the Complaint The patient may be sufficiently aware of these isorders of memory to complain of them or they may have been noticed by others who will seek the physician in behalf of the patient

Functions of Affectivity or Feeling THE PERVASIVENESS OF AFFECT The ions of affectivity or feeling consist of the psychic phenomena known as feeling ones moods and emotions Feeling tones accompany all sensations and ideas and e tinged by either pleasure or displeasure both of which are conducive to states or emotions that may be termed moods Moods usually are prolonged states char cterizing persons as having for instance a sunny disposition or a grouchy disposi i Moods accompany all of the episodic emotional states of delight disgust joy ief love hatred anxiety peace anger and their modifications

Pleasurable Affects Euphoria is a pleasurable affect The individual is imbued ith a subjectively pleasant feeling of well being Euphoria is most frequently ob erved in general paresis multiple sclerosis and in some instances of frontal lobe mor Euphoria may readily shift to irritability Euphoria is contrasted with an air enjoyment and of self-confidence which radiates from some normal individuals

A mood of entrancing peaceful rapture and tranquil sense of power is termed *stasy* A religious element is an essential part of the state because it permits of etachment from worldly things and the religious tenet of rebirth and thus purifica E Ecstasy represents the achievement of the maximum of wish fulfillment The tion is observed in hysterics epileptics and schizophrenics

Depression An affective dejection varying from mild downheartedness to stupor is known as depression A reactive depression arises from sickness bereavement betrayal loss of property job or money and from many other difficulties Autonomous depressions arise from unrecognized affective factors the sources of which are beyond the patient s capacity for understanding

Tension Tension is characterized by a feeling of uneasiness restlessness dissatisfaction and expectancy It is really a manifestation of anxiety

Fright Fear Anxiety and Panic Fear and fright are mechanisms intended for the preservation of life They are therefore comparable to pain It is possible to abolish pain and fear temporarily and to a fairly predictable degree with certain drugs

There are no essential distinguishing features between fear and anxiety When the threat of danger is acute fright seems to be the preferable term When a continuous danger impends it is proper to designate the reaction as one of anxiety

Of fear in children and primitive man four aspects can be distinguished

1 The *mimic* or *pilomotor reflex* is characterized by changes in the skin which especially in animals lead to a sort of masquerade In man a lively reaction takes place in the skin under the influence of serious emotion There may be shakes and vasomotor changes and the face loses color There is a feeling of intense cold In some an unbearable itch and urticaria make an appearance The skin may desquamate after a few days Spontaneous extravasations occur in face and legs painful hypersensitivity of the skin neuralgic pains general hyperesthesia of the senses of perception and complaints of cold and pain may be present and chronic rheumatism may flare up

2 The *flight reflex* is expressed by mobilization of every facility to escape the danger There is an accelerated activity of the sympathetic system with increased supply of epinephrine to the tissues increased blood supply halting of motility and secretion of the digestive organs or in some cases hyperfunction such as passage of bowel movements Reactions of the vascular system are prominent Cardiac palpitations and sudden sthenocardiac pressure are well known A weak heart may not survive The common distress reactions are gagging anorexia nausea and diarrhea Polyuria and involuntary urination are common A latent exophthalmic goiter may be made active Impotence a feeling of paralyzed sexual functions alternating drowsiness and sleeplessness often ensue

3 A sudden stop to this aggressive activity is *panic* There are a psychic stand against the threatened danger and a somatic mobilization All psychophysical possessions are geared for defense

4 The popular expression I was literally scared to death may actually be termed the *death reflex* as the clutching of objects in the hand at the time of drowning There is a general paralysis of functions and cataleptic posture and a stiffening due to fear are present Cataleptic stiffening after great fright is often experienced In some cases disease symptoms which resemble parkinsonism anxious eyes (proptosis) tremor of the hands stiffening of muscles (a sort of decerebrating stiffness) ensue

The breathless stiffening of fright may cause in some an attack of asthma a fear to breathe not influenced by any therapy A chronic nervous asthma may result There may ensue a drop in weight from loss of fluid due to profuse perspiration and internal secretory changes Paresthesia of the extremities hands and feet going to sleep and acrocyanosis may ensue

Fear whether of magic evil spirits or the dark is present in most men When resistance is lowered by disease by shock or by psychic conflict fear may become rampant Depending on early training of the individual fear may develop of dark strange places loud noises high places lightning thunder and the graveyard on a bright moonlight night

Inadequate Affect This is emotional dulling characterized by a lack of sensitiveness to those experiences that normally give emotional response The qualities of gratitude sympathy hope anticipation grief regret pride or shame are partially or completely lost

Depersonalization This is characterized by a feeling of unreality and of changed personality It is a form of withdrawal from reality The patient feels that he is no longer himself but he does not feel that he has become someone else There is a total loss of affective response everything seems unreal Depersonalization occurs in depressions in anxiety obsessional states hysteria and in some schizophrenias It is more frequent in intelligent sensitive affectionate introverted and imaginative types of individuals than in other types

Relationship to the Complaint Evidence of abnormality of the affect (feeling tones moods or emotions) if present is very likely to appear in the patient s complaint

Conative or Striving Functions The conative or striving functions of the human being relate to volition or will and action and timed behavior The normal person strives to attain certain goals The basic or natural aims of this striving are self preservation race continuation acquisition of knowledge and its dissemination social intercourse and the appreciation of beauty right conduct and all the will for the good things of life

Attitude is an acquired conscious desire or an unconscious predisposition to react in a characteristic manner toward a given type of person or situation

Disposition is a sum of the individual s tendencies as determined by the affective and conative components of his personality

Aggression is the deep seated drive or pattern of the personality possessed by an individual who reacts in a positively forceful way Aggression carries the implication of will to power and too of hostility and attack These tendencies may or may not be obviously expressed In the desirable individual the aggressive impulses are sublimated so as to serve to promote constructive forms of unselfish adaptation

Ambivalence implies a combination of the use of the action sphere and the affective sphere of the personality In manifest ambivalence the individual may be abnormally impelled by contrary drives

Disturbance of activity comprises states of (1) hyperactivity or overactivity (2) decreased activity (3) repetitious activities (4) automatic obedience (5) negation and (6) compulsion

In *hyperactivity* the patient is very busy but the activities often are not productive The stream of talk expresses a flight of ideas In *decreased activity* there is delay in starting an activity and once begun it is executed slowly *Repetitious activities* are variously expressed for instance a constant repetition of an activity is *stereotypy* a maintained immobility of position is *catalepsy* A cataleptic form of immobility is frequently observed in schizophrenics and it is termed *waxy flexibility* Schizophrenics perform many different sorts of mannerisms such as grimaces repeated gestures and peculiarities of gait Comparable repetitious activity in speech is observed A reiteration of speech is known as *verbigeration*

Automatic obedience may be expressed by command automatism in which suggestions or requests are compulsively followed There may be a repeating of words or phrases which is termed *echolalia* When words phrases or movements are imitated without command these acts are termed *echopraxia*

Negativism is a refusal on the part of the patient to do what is desired of him or is the performance of the opposite Extreme negativism is characterized by mutism refusal of food noncompliance with requests and resistiveness

It may be a difficulty with the conative or striving function which causes the patient to seek the help of the physician If so at least a hint of difficulty may be found in the complaint which the patient makes

The Manic Depressive Group of Psychoses This is a group of psychoses characterized by manic states (manias) by depressive states (melancholias) or by mixed manic-depressive states In some cases the patients suffer from only one attack either of mania or of melancholia not infrequently there is a regular recurrence (cycle) of the two opposite states of mania and melancholia the single cycles repeating themselves after shorter or longer free intervals throughout the whole lifetime—cyclothymia

THE MANIC PHASE In the manic phase of this psychosis the patients experience and manifest an exalted mood they tend to sudden changes in humor to flight of ideas to easy distraction of the attention In the exaltation everything is easy for the patient The facial expression is happy though subject to rapid change to anger There are rapid changes in the direction of the ideas the patient seems unable to hold an idea in mind objects about him quickly altering the flow of ideas There is a tendency to make rhymes puns and alliterations The pressure of activity exhibits itself as a preternatural loquacity or in a tendency to excessive busyness in the *writing of many letters in the sending of many telegrams in the making of unnecessary* purchases in unnecessary traveling or visiting Such patients are prone to indulge in sexual and alcoholic excesses

Mild forms of mania sometimes go unrecognized The patients are often thought to be unusually healthy and vigorous Once the patients are under observation the superficiality of it all is easily recognized the poverty of the underlying thought the tendency of the mental associations to be influenced by sound similarity and contiguity the resemblance to mild alcoholic intoxication and the absence of insight into the disease

In the severer forms of mania there may be delusions of grandeur and of extravagant self appreciation The mood changes suddenly without warning there may be *an increase of irritability or an outbreak of anger and violence The patient may* attack the surroundings and explode into blackguardism Patients of this type gesticulate dance yell tear up their clothes or pull out their hair In the severe forms of the disease they may eat drink or smear themselves with urine or feces Under all this maniacal excitement they experience no feeling of fatigue

In milder psychoses the orientation may be good but in the severer forms especially those with many hallucinations there are disorientation and confusion

The patients lose weight rapidly from insomnia lack of food and excessive movement After several months during which remissions and exacerbations occur the patients may gradually improve regain their weight and return approximately to normal

The manic states have a marked tendency to recur The first attack usually appears in youth

A maniacal attack of the manic depressive group must be distinguished from (1) the maniacal attacks in dementia paralytica (2) catatonic states of schizophrenia and (3) hallucinatory confusion or amentia

THE DEPRESSIVE PHASE The depressive phase or melancholic state of the manic depressive psychosis is characterized by a pathologic sadness and a slowing of thought and emotion associated with ideas of self-depreciation ideas of poverty ideas of sin and hypochondriacal ideas There is always danger of suicide Digestion is disturbed and the bowels are constipated In the milder states the patients *admit* that they are sick in the severe states they deny that they are sick and may assert that they are just sinful

A large part of the involutional psychoses (senile presenile and climacteric psychoses) take the form of melancholia

The psychomotor retardation is the clue which distinguishes depressive phases of the manic-depressive psychosis from other forms of depression The depressive phases of the manic-depressive psychosis must be differentiated from (1) depressive

phases of dementia paralytica (2) depressive catatonic states and (3) depressive stages of senile dementia

MIXED MANIC AND DEPRESSIVE STATES Not only are depressive symptoms prone to alternate with symptoms of exaltation but states are met with in which some of the manic symptoms are combined with some of the depressive symptoms

RELATIONSHIP TO THE COMPLAINT The patient s manner and speech when he makes his complaint often betoken one or the other of the states described in great or slight degree

The Paranoid States By a paranoid state is meant a form of mental disturb ance in which delusions often systematized in character and especially delusions of persecution and of grandeur develop with or without hallucinations

A paranoid state may develop acutely or it may develop and grow into a system with progressive increase in delusions

The acute paranoid states are common in chronic alcoholism taking especially the form of the acute jealousy insanity A similar jealousy insanity sometimes de velops in the puerperium or during lactation

In the chronic paranoid states there are anomalies both of the intellect and of the emotions The disease develops so slowly that for many years a psychosis may not be suspected The patient is often hypochondriacal retiring and suspicious and suffers from ideas of reference Later ideas of persecution appear such as that people treat him badly have it in for him and are trying to poison him Egocentric grandiose ideas usually develop such as ideas of unusual personality and powers revelations from God royal descent ideas of reform and of discovery In some cases hallucinations voices and visions are prominent

According to the nature of the delusions the paranoid states are described as erotic mania religious mania political mania and jealousy mania Paranoiacs are often homicidal

Most cases can be placed in one of two groups (1) true chronic paranoia and (2) dementia paranoides In the latter the course is much quicker the delusions are more nonsensical and a high grade of confusion and dementia may soon appear

If the patient complains of being persecuted or if any of the other characteristics mentioned are evident in what the patient complains of the physician should suspect the existence of a paranoid state—but he should not jump to the diagnosis

The Schizophrenic Psychoses (*Dementia Praecox*) The schizophrenic psychoses are often termed the psychoses of adolescence (dementia praecox) a group of mental anomalies presenting marked individual variations but all having in com mon (1) development about the time of puberty or at any rate in the first half of life and (2) termination sooner or later in a dementing process of variable grade Not all reach a high grade of dementia in some cases there is arrest with defect in a very few cases there may be complete recovery The cause is unknown

In diagnosis the episodic states are less important than (1) the motor symptoms and (2) the development of a progressive dementia in youth

Among the variable clinical divisions of these adolescent psychoses three main types may be distinguished (1) hebephrenia (2) catatonia and (3) dementia paranoides

HEBEPHRENIA An enterprising youth at puberty or soon after begins to stand still or to change his occupation frequently has an exaggerated idea of his own importance tends to occupy himself with the deepest problems of existence shows pleasure in resounding phrases and dry proverbs or abnormally devotes himself to puns and to practical jokes Signs of feeble judgment soon appear Victims of this disorder write long theoretical treatises publish impossible books or undertake im practical careers Among the complicating episodic states may be mentioned sudden depression excitation anxiety or suicidal tendency Each episodic state is followed by deterioration and by increase of apathy

The disease usually goes unrecognized for some time and is designated neurasthenia or hypochondriasis Hebephrenia should be suspected in young persons with good memory and comprehension who exhibit loss of initiative, dulling of the higher feelings or a sense of failure of intelligence particularly when no special cause for exhaustion is demonstrable and when the symptoms are not easily got rid of by general hygienic measures

CATATONIA One of the commonest forms of insanity in young persons is that characterized by (1) specific catatonic phenomena such as negativism command automatism stereotypy grimacing anomalies of speech and writing (2) episodic states of stupor or excitation and (3) a termination in dementia

After a brief period of mental depression at the onset the patients suddenly begin to behave in a surprising manner Without apparent cause they may suddenly refuse to take food or they may remain away from home without cause or they may enter upon some impossible marriage engagement Sooner or later symptoms of catatonic excitation develop In a few cases the patient attempts or commits suicide early in the disease the family not having suspected the existence of a psychosis

In catatonic stupor there is a peculiar alteration of psychomotor innervation including negativism in which the patient shows resistance (1) to any external influence on his will and (2) to spontaneous motor impulses arising in himself manifested by refusal to speak to show the tongue to take food to swallow saliva Muscles once innervated may retain their innervation giving rise to the so-called stereotyped attitudes such as inclined head crossed legs snoutlike projection of mouth and turned in thumbs These attitudes may be maintained for hours days or weeks despite discomfort or ulceration from pressure

In command automatism on the contrary there is in contrast with negativism a preternatural suggestibility in the motor domain exhibited as waxy flexibility echolalia and echopraxia

In catatonic excitation violent purposeless movements are made often with loud cries and attacks on surrounding people sometimes there are suicidal attempts The so-called monotonous or stereotyped movements such as rhythmic swaying of body rubbing of hands movements of tongue are kept up for hours or days at a time and represent a transition stage between catatonic stupor and catatonic excitation

In intervals between excitation and stupor certain peculiarities of behavior usually remain such as grimacing motiveless laughing or crying coprophagy peculiar writing tendency to verbigeration irrelevance and mannerisms On the psychic side there are (1) a slow enfeeblement of the judgment though the recording faculty and the memory may be good (2) a dulling of feeling and apathy (3) sometimes delusions and (4) an incalculability and bizarreness of behavior

Certain somatic signs may coexist exaggerated reflexes heightened mechanical excitability of muscles and nerves increased secretion of sweat sebum and saliva amenorrhea subnormal temperature and remarkable variations in body weight Remissions and exacerbations are common

The catatonic phenomena alone do not suffice for the diagnosis of schizophrenia for they are met with temporarily in other psychoses The combination with mental enfeeblement is essential The differentiation from dementia paralytica, epilepsy hysterical twilight states or amentia is usually easy In imbecility and idiocy the patients have been intellectually feeble from early childhood whereas in dementia praecox there is a demonstrable deterioration of the mental powers The differentiation from a manic depressive psychosis is not always easy but the apathy the negativism and the evidences of actual mental deterioration will usually be decisive

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Acute Hallucinatory Confusion (Amentia Acute Confusional Insanity, Acute Hallucinosis) In various infections intoxications and states of exhaustion especially after surgical operations an abnormal mental state may appear characterized by (1) the sudden onset of dreamy clouding of consciousness (2) numerous and very lively disconnected hallucinations and delusions and (3) peculiar motor symptoms consisting either of increased impulse to movement or of stuporous inhibition The patients are confused disoriented and incoherent the recording faculty is markedly disturbed the mood is anxious and variable Some of the psychoses of pregnancy and of the puerperal period belong here The patients usually emaciate rapidly and they suffer from anemia anorexia and insomnia

Amentia usually lasts from 6 to 9 months or even longer than a year Many die from inanition or from complicating infections some commit suicide about one third recover a few go over into chronic mental disease

Acute hallucinatory confusion is to be distinguished from (1) manic depressive states (2) epileptic insanity (3) catatonic form of dementia praecox and (4) dementia paralytica

Relationship to the Complaint These manifestations are likely to be unrelated to the complaint which the patient originally made but may be related to those of a relative or guardian of the patient when the physician is called to meet a frightening emergency

Psychoneurosis Without a clear conception of the meaning of the complaint a practitioner of any form of the healing arts cannot fulfill the first obligation to the patient This obligation is a correct diagnosis In order to evaluate and assign a specific connotation to many a complaint or complaints a wide familiarity with organic and psychogenic illness is required For it must not be forgotten that a psychoneurotic illness renders no immunity to organic illness and likewise an organically ill patient may have and may have always had a psychoneurosis The psychoneurosis may cause a very great difficulty in the interpretation of the complaint for the somatic and the psychologic components in any given complaint must be disentangled Until the somatic and the psychologic components have been separated it may not be possible to determine whether an illness is primarily or wholly psychogenic or whether it is somatic with coexisting psychogenic factors

Emotional factors play a predominant role in the production of a psychoneurosis These factors usually have begun to operate in early life and often are manifested by such disturbances as sleep walking crying out in sleep enuresis disturbances of speech food idiosyncrasies delirium accompanying slight rises in temperature de*structiveness emotional* excitability tantrums phobias *compulsions shyness nail* biting and other behavior affections

In the psychoneuroses the personality remains socially organized It does not deny the existence of reality it attempts to ignore it The interpretations of the environment remain unchanged although certain elements are given abnormal affective values Repression is maintained but the repressed material returns in a distorted form so that it is acceptable only to the patient

It seems that many persons are unusually sensitive to the tension and conflicts of life and react to these with faulty reactions which impair the efficiency of the individual These faulty reactions often create anxiety which is one of the commonest and most important sources of psychoneurotic disorders Anxiety disturbs physiologic functions In acute forms anxiety may produce spasm of the cardiac and pyloric portions of the stomach intestinal spasms hyperchlorhydria diarrhea or constipation palpitation tachycardia extrasystoles vasomotor flushing and increase in body temperature (99 to 100 F) and respiratory distress The hands and face perspire The patient may have fidgety movements of hands or feet the voice may be uneven or strained and the pupils may be widely dilated Anxiety therefore

DEMENTIA PARANOIDES Dementia paranoides is an adolescent psychosis which is observed less commonly than either hebephrenia or catatonia It is characterized by a rapidly advancing delusional state in which the delusions quickly become more surprising and nonsensical terminating relatively quickly or in a few years in feeble mindedness the symptoms are changeable during its development and course and are characterizable by episodic states of depression and excitation

At the onset depressive symptoms usually predominate (insomnia self accusation blueness unrest suspicion sense of change in the environment) and ideas of reference prevail Illusions hallucinations and delusions soon appear the latter consisting chiefly of grandiose or of persecutory ideas The delusions multiply rapidly though the patient s recording faculty and orientation remain little disturbed The gross irrationality of the delusions is characteristic suggesting that of dementia paralytica Delusional falsifications of memory are common the patient tells stories of marvellous experiences or of wonderful personal acts

There is no disease insight The apathy and indifference are characteristic The dementia progresses rapidly with the appearance of incoherence in speech and writing

The diagnosis may be difficult at first but is easy later on The condition must be differentiated from (1) chronic hallucinatory paranoia and (2) dementia paralytica The age somatic signs and findings in the cerebrospinal fluid will eliminate syphilis of the nervous system

RELATIONSHIP TO THE COMPLAINT The complaint made by a victim of schizophrenia is likely to be confusing or unenlightening but a hint given by the patient may lead the physician to identification of diagnostic features in the history or signs

The Infectious Toxic and Exhaustion Psychoses Under this heading are listed the initial deliria febrile deliria and convalescent deliria of the infections and intoxications In infectious diseases delirium may appear at the beginning or during the main course or in convalescence These forms of delirium may occur in the course of any infectious disease There need be no relation between the height of the fever and the intensity of the psychic symptoms

FEBRILE DELIRIA In the febrile deliria the principal symptoms are (1) clouding of consciousness (2) disorientation (3) hallucinations in addition there may be (4) violent motor excitement sometimes resembling that of mania or (5) stuporous states

DELIRIA OF CONVALESCENCE The deliria of convalescence may appear suddenly after the temperature has become normal They consist of (1) dreamlike confusion (2) hallucinations and illusions (3) occasionally delusions and (4) lively motor excitement The patients are usually anxious and suffer from self accusation ideas of sin and other micromanic delusions sometimes instead the state may be euphoric Stupor and excitation may alternate The outcome of these deliria is usually favorable

COMA SEMICOMA STUPOR AND CONFUSION *Coma* is a state of complete unresponsiveness to the environment The patient s reactions are limited to elemental reflexes and even some of these are impaired *Semicoma* is a condition in which there is responsiveness to some of the elemental mental functions such as calls by name reacting to noise and reacting to a visual threat There is no significant defect in the elemental reflexes *Stupor* is a condition of greater awareness than semicoma in which the patient has no defect in elemental reflexes responds to elemental mental functions and in addition reacts to verbal commands having an ability to execute simple things such as shutting the eyes and sticking out the tongue *Confusion* is a state in which there is only a mild disturbance in the state of consciousness The elemental reflexes elemental mental functions and reactions to simple commands are usually intact These patients present chiefly orientation difficulties

in lies They do not intentionally lie this lying is but the pathologic phenomenon due to faulty reproduction This faulty reproduction or transformation of memories is the so-called pseudologia fantastica or mythomania These patients may have a complete but transient amnesia or the amnesia may concern only persons or events

The feelings of the hysterical individuals are easily hurt These individuals believe that they are misunderstood they think that others do not pay them the attention that is their due or that others intentionally slight them

Many hysterical patients may be unable to eat or they vomit almost constantly a condition termed anorexia nervosa In others the abdomen may be continuously distended with gas and this is termed hysterical meteorism or the patient is called a bloater In a few patients there may be a retention of the urine A rare form of the disease is pseudocyesis the patient even without exposure may think herself pregnant

There may be transitory disturbances of consciousness such as hysterical twilight states somnambulisms or trances The hysterical twilight states appear suddenly There is confusion and silly childish behavior resembling simulation and the condition may last for hours days or weeks

The hysterical trances or lethargic states may be characterized by sleep for days weeks or months though enough consciousness may be maintained to enable eating drinking voiding and defecating If aphasia is present the patient may not have any difficulty in communicating in whispers The hysterical mute can communicate freely and correctly by writing

Standing and walking may be difficult or impossible for the hysterical patient When the patient lies down no disturbance of motility sensibility or co-ordination can be made out Some patients who cannot stand or walk can crawl or can swim The condition is commoner in women than in men and in young people than in old It was formerly termed astasia abasia There may arise a complete immobility on account of pain although no cause for the pain is discoverable The symptom is an hallucination in the pain sense Some of these patients during the attacks of pain manifest tachypnea and tachycardia

The Ganser syndrome is a paralogia It has been frequently present among prisoners of war awaiting trial As in many other hysterical states the hysterical protection from a full realization of the true situation affords a relative freedom from the anxiety which might otherwise be intolerable

The recognition of hysteria is usually easy through the characteristic stigmas and through the exaggerated often theatrical character of the mental symptoms and the capricious oscillatory course A hysterical patient also can have an organic disease

Accident Neurosis (*Hysteria*) In persons of a paranoid tendency those who are insecure those who crave sympathy and attention and those continuing work beyond mental or physical capacity unpaid financial obligations or the failure to derive satisfaction from work all may predispose to a traumatic neurosis The conviction on the part of the patient that there is the right to expect compensation may be a motivating factor A period of time between the injury and the appearance of the mentally determined symptoms which is occupied with vague affective disorders often supervenes

The symptoms of the accident neuroses except in the matter of compensation differ in no essential feature from those of other neuroses In those cases in which compensation is a participating motive the patient remains unaware of such an influence and denies the desire for compensation Underlying feelings of guilt may be overcompensated and experienced as self pity

Occupation Neurosis In occupation neuroses the patient on attempting to execute some specialized movement performed in an occupation requiring the co ordination of groups of muscles suffers from a spasm of the muscles involved in

together with the various mechanisms developed to avoid it constitutes an important factor in the development of psychoneurosis

The psychoneuroses are said to be more frequent in women than in men This is probably a matter of degree and sex of the author rather than of incidence Therefore it is not well in diagnosis to infer that the numbers of psychoneurotic women greatly exceed those of psychoneurotic men for they do not and there is no reason that such should be true

Most neuroses of adults develop between late adolescence and 35 years of age This is the period when the individual becomes confronted with the problems of adult adjustments and responsibilities It is the period when satisfying social economic and sexual situations should be established Frustrations in these fields may lead to conflict anxiety and tension that are met only by neurotic mechanisms

Hysteria Hysteria is characterized by an abnormal mental state in which there is a disproportion between affective reactions and the stimuli that give rise to them The suggestibility of the patient is heightened the sudden transformations of the emotional states and the moods are best described by a marked degree of caprice

The disease is much commoner in women than in men It begins in adolescence sometimes in childhood Most patients have a distinct infantilism in the psychopathic field Provocative causes are fright, anxiety and trauma

Hysteria may begin with a feeling of constriction in the neck or as a lump in the throat which cannot be swallowed usually associated with loss of the pharyngeal reflex and clavus hystericus or pain in the region of the large fontanel described as resembling a nail being driven into the head Hysterical paralyses include hemiplegia monoplegia aphonia astasia abasia ptosis and retention of urine Hysterical blindness and agnosia may be present in some instances

Hysterical hyperesthesias include the so called spinal irritation or tenderness on tapping the spinal column ovarian tenderness on pressure in the region of the ovary and various hyperesthetic zones occurring in any part of the body Vasomotor and secretory lability is often expressed in hot and cold flushes and localized disturbances of sweating

Hysterical convulsive seizures occur in which patients without injuring themselves sink down and assume and maintain for some time remarkable attitudes such as the arc of a circle or various pathetic or theatrical positions or exhibit wild rotating movements The attack can be terminated by a dash of cold water The more common hysterical seizures include silly laughing crying coughing hiccough torticollis and muscular cramps

Hysterical anesthesia may affect one extremity one half of the body or two extremities simultaneously becoming ever more marked the more attention it receives Often the anesthesia has a topographic distribution corresponding to areas of skin covered by certain articles of clothing such as the gloves chemise drawers or stockings Exaggerated reflexes with false ankle clonus may be present

The principal mental stigmas are a pathologic lability of mood fallacies of memory ideas of reference and lack of consideration for others

The hysterical patient is abnormally irritable slight annoyance may lead to explosive behavior and thus the expression he became hysterical It sometimes seems as though the smallest stimulus excites the greatest reaction The smallest and seemingly most insignificant incidents may give rise to convulsions and twilight states However in an emergency the hysteriac may exercise a great calmness of action The hysterical patient may be happy jolly and easily accessible one moment and the next moment without any apparent reason become the reverse—irritable suspicious and sullen This pathologic irritability is more intense at the menstrual periods at the menopause and during pregnancy than at other times

Hysterical patients are incapable of objective report Often they seem unable to distinguish what they remember from what they imagine They are thus often caught

Since the patient is perfectly sane his complaint often is thoroughly descriptive

Neurasthenia Psychiatrists believe that the cause of neurasthenia is mental and they no longer believe that a generally impaired or exhausted physical state in which the nervous system is drained of its energy is ever the cause of this disorder In general diagnosis and the general practice of medicine a greater number of patients will be found who are physically exhausted and who do recover from the so-called neurasthenia after rest and success than the number whose symptoms are due to a purely mental disorder such as repressed hostility constant failure frustration disappointment boredom and monotony

In neurasthenia there are a sense of mental and physical fatigability tension irritability painful and other abnormal sensations and depressive affective tendencies There may be complaints of dizziness of a feeling of pressure on the head and perhaps of pain in the back of the neck Often there is an intolerance of noise bright lights and cold Gastrointestinal complaints such as gas indigestion constipation and diarrhea are common Often the skin is flushed and sweating In men an impaired potency and failures at intercourse may occur Advancing age not neuras *thenia is the usual cause of failures at intercourse in men who are more than 50* years old

In the morning the patient awakes with a feeling of exhaustion but as the day progresses there is improvement and by evening there is comparative freedom from the exhaustion Many patients are shy and awkward and lack confidence in themselves They may be critical dissatisfied envious and resentful and may appear to take pleasure in finding fault with and annoying others

Psychoneuroses usually if not always show mixed features It is most important to distinguish neurasthenia from the depressive form of manic depressive psychosis and early schizophrenia These differentiations are made by the psychiatrist through time and observation

RELATIONSHIPS TO THE COMPLAINT The complaint voiced by the patient often is of diagnostic importance

Hypochondriasis Hypochondriasis represents a faulty type of adaptation to personality inadequacies with which a sense of disappointment or of guilt is associated The presenting symptom is an insistent protracted preoccupation with organs or bodily functions The hypochondriac is aware of various sensations which most persons do not notice These various sensations are magnified out of all reasonable proportions The patient complains of these things

Pain Pain is of primary concern in most complaints The evaluation of pain in the complaint aids in diagnosis and management of the patient Pain is a sensation like smell taste or vision It can be further compared to these sensations for it is first a sensory perception This sensory perception is the pain threshold Pain has a reaction which is usually unpleasant but not always so However the important conception of diagnostic value of pain is the fact that sweating tachycardia digestive disturbances anxiety fear terror panic and prostration may be solely reactions to pain

Pain is a centrally integrated experience the painful sensations being derived from impulses traversing specific pathways According to Wolff and Hardy all fibers carrying impulses experienced as pain enter the spinal cord through the dorsal root ganglions After entering the cord these impulses are conveyed across to the opposite side where the pathways are localized in the anterolateral portion of the spinal cord The fibers of the spinothalamic tract pass into the nucleus centralis posterior of the thalamus The cortical projection from the nucleus centralis posterior is predominantly to the postcentral convolution It is probable according to Wolff and Hardy that the brain structures involved in pain perception occur in both hemispheres in the region of the central fissure

The dual aspect of pain that is the distinction between perception and reaction

this act The function of the muscles remains unimpaired when the same muscles are employed for the performance of some other act

RELATIONSHIP TO THE COMPLAINT In the complaint which the patient makes he often reveals to the physician the hysterical nature of his underlying condition

Psychasthenia (*Compulsion Neuroses*) Psychasthenia includes phobias, obsessive thinking and compulsive acts

A *phobia* is a fear or dread associated with an idea object or situation Among the common phobias are fear of dirt of bacteria or of certain animals To many of these ideas objects or situations Greek prefixes are applied and thus are produced such terms as claustrophobia, the fear of confined spaces The phobic patient cannot avoid these fears and if forced into an anxiety producing situation a state of agitation follows the patient cries and shakes all over may have general tremors and be unable to continue the duty at hand, and may become overwhelmed with panic

A phobia is often a fear of disease Reassurances that somatic disease is not present often accomplish much in some patients In others such assurance is of no value

Obsessive thinking is defensive in purpose Distressing thoughts are composed of contents repugnant to the individual's conscious moral and esthetic feelings In a mild degree these are normal for everyone has such thoughts for instance of tipping over a glass of water on the dinner table when too much formality is being practiced about the table

Obsessive thinking characterized by persistent doubting vacillation and indecision may assume the form of *folie du doute*

Compulsive acts are characterized by an irresistible urge to perform a certain repetitive stereotyped act One of the commonest compulsive acts is that of getting out of bed after retiring to check the gas or electricity or to see if the doors are locked The compulsive act reduces the intensity of anxiety

Since everyone has defensive character traits the diagnostic decision must often be arbitrary In the fully developed cases with the well marked obsessive thinking fears or compulsive acts there is of course no question

A failure on the part of the patient to regard his phobias and compulsions as absurd indicates a schizophrenic origin

The depressions of manic depressive psychosis may be accompanied by obsessive ideas

Anxiety has been defined as a state of heightened tension often accompanied by somatic reactions usually in the vasomotor sphere It may arise when there is a threat to the personality Anxiety often arises in association with frustrations related to property job money sex or marital adjustment The patient usually recognizes the tension and anxiety producing agent and often there is nothing that can be done about it

The anxiety is often expressed by symptoms of depression irritability restlessness psychosomatic disturbances outbursts of aggressiveness attacks of weeping and feelings of inadequacy and inferiority accompanied perhaps by a paranoid attitude There may be difficulty in falling asleep disturbance by fearful dreams suffering from coarse tremors or trembling The hands and feet are cold and clammy The patients are absent minded There may be periodic acute panic like exacerbations lasting an hour or two These acute attacks take many different forms of representation such as rapid heart action precordial discomfort nausea diarrhea desire to urinate dyspnea and a feeling of choking or suffocation The pupils are dilated the face is flushed the skin perspires the patient suffers from paresthesias and tremulousness feels dizzy or faint and often has a sense of weakness and of impending death The patient may express beseeching and apprehensive appeals for help

sensory peripheral nerve fiber or a nerve trunk be injured the threshold for pain is altered or obliterated In the central nervous system the pain threshold is altered or obliterated by spinal cord injuries Lesions in or near the internal capsule of the brain may elevate the pain threshold

The most important alterations of the pain threshold to be evaluated in the complaint are those resulting from psychologic factors These factors may raise the pain threshold for instance during intense emotion as during fright severe injuries may be sustained without knowledge Autosuggestion hypnosis distractions malingering and mental concentrations all raise the pain threshold Anxiety tension and fear seem to lower the pain threshold A lowered pain threshold is never solely due to structural disorder of the nervous system

THE LEVELS OF PAIN INTEGRATION IN THE CENTRAL NERVOUS SYSTEM There are three levels in the central nervous system at which pain may be integrated the tectum mesencephali the thalamus and the cerebral cortex

In the tectum mesencephali the appreciation of pain is no more than a feeling tone and therefore probably is closely related anatomically and physiologically with the activity of the hypothalamus its powers to perceive pain having been taken over during the process of evolution by the thalamus

The fact that pain is relatively little influenced by small cortical lesions is the result of the numerous pathways which are provided by the thalamus for the passage of pain sensation to the cortex

The appreciation of pain stimuli at the cortical level is modified by the state of activity of other cortical centers so that the perception may be aggravated in some instances and prohibited in others

Wolf has named six basic mechanisms which are responsible for headaches of *intracranial* origin These are (1) traction on and displacement of the great venous sinuses or traction on the veins that pass to the venous sinuses from the surface of the brain (2) traction on the middle meningeal arteries (3) traction on the large arteries at the base of the brain (4) distention and dilatation of intracranial arteries (5) inflammation in or about any pain sensitive structure of the head and (6) direct pressure by tumors on the cranial nerves containing pain afferent fibers

The *headache associated with the fever* of acute infections is due primarily to the distention of the intracranial arteries The spontaneous increase and decrease of intensity of the headache of fever parallel the changes in amplitude of pulsations in these arteries The amplitude of pulsation of the cerebrospinal fluid in the headache of fever is greatly increased during the fever These amplitudes of pulsation of the cerebrospinal fluid during fever headache are in marked contrast to the amplitude of pulsation in migraine which is not accompanied by such a disturbance It may be that these pulsations account for throbbing headaches of fever

Headache from intracranial hypertension or disease is usually referred pain from the affected intracranial pressure Pain referred to the head from disease of tissue not in the head does not occur Fever is often accompanied by headache but this is not referred pain

Headache is a common early symptom of a space occupying lesion within the skull sometimes it is the initial symptom The absence of a headache however does not preclude the possibility of a space-occupying intracranial lesion The symptom headache as arising from intracranial pressure always requires assessment

The headache from a space occupying lesion intracranial hypertension renders the sufferer clearly intensely preoccupied with pain and the questions and physical examination are but an addition to the burden of the pain These patients observe an economy of movement and of speech since effort aggravates the pain The pain is never spoken of as continuous over long periods since organically determined headache is never so continuous but always is subject to intermission over varying

to pain is apparent and is easily appreciated in the case of heat light, touch cold and olfactory perception The reaction pattern of the organism to noxious stimuli has man[illegible] components Reaction to pain is modified by conditioning experiences such as [illegible]tement of a game or combat the apathy that accompanies tissue damage [illegible] certain religious and mystic practices and the indifference to tissue damage during sexual excitement

Total absence of the usual physiologic behavioral responses to noxious stimuli is rare Ford and Wilkins described three children with lack of reaction to pain rather than absence of pain as sensation They could appreciate light touch and could distinguish between warm and cool test tubes and between the head and the point of a pin They were however indifferent to damaging stimuli Kunkle and Chapman described a young man who had almost complete insensitivity to pain and attacks of unconsciousness beginning in childhood Boyd and Nie described an otherwise apparently normal girl 7 years old who was completely insensitive to pain The usual painful responses could not be obtained by any stimulation She was able to distinguish sharpness from dullness and the senses of touch vibration, position and agnosis were normal Boyd and Nie suggested two hypotheses to explain the patient s indifference to pain (1) There exists a congenital structural defect with incomplete neural connections and communicating fibers in the post central area with consequent inability to organize the complex concept of pain (2) There is no neurologic defect but there is indifference to pain which represents an aphasia like disturbance

McMurray reported an exhaustive study of a patient with universal insensitivity to pain At no time during any of the experiments in which noxious stimuli were used did the patient report a sensation or feeling that could be interpreted as a report of pain Consistent with this was the complete absence of any observable signs of painful experience

Varieties of Pain Two varieties of pain are recognized superficial and deep Superficial pain is characterized by the quality of localization and has the components of sensations described as pricking sharp burning or itching it incites the action of fight or flight Deep pain possesses the qualities of a diffuse aching or of a colic It incites the actions of withdrawal forces a cessation of activity and causes the affected one to seek protection Deep severe pain may cause nausea and vomiting

Threshold of Pain *Pain perception* is the threshold of the amount of stimulus required first to produce a painful sensation In superficial pain the pain threshold can be elicited by heat pinprick or chemical agents The threshold for deep pain can be elicited by electric or mechanical means such as a faradic current or the stretching of a hollow viscus The determination of pain threshold experimentally as well as in practice is based solely on the accuracy of the verbal report Despite the inaccuracies of the verbal report the pain threshold in man is relatively stable and uniform The threshold for the perception of pain is found to be of the same order of uniformity as is the pulse rate the number of leukocytes of the blood or the blood pressure The difference of pain threshold in different individuals lies in the individual reactions to pain Generally it may be asserted that the alarming experiences aroused in the mind of the patient are the most distressing aspects of pain For instance morphine does not discontinue the perception of pain but it does remove the fight flight anxiety reaction of the sufferer

The pain threshold like the other somatic components may be altered The pain threshold is lowered by traumatic deformation denudement or injury of tissues near the sensory nerve endings responsible for the reception of pain stimuli For instance a mildly sunburned or a frost bitten skin is twice as sensitive as a normal skin The threshold for pain is raised or obliterated by local anesthetization If a

then is carried in this neuronic pathway to the brain and is perceived as coming from the distribution area of the second neuron

The most numerous and important of reflected pains are those due to pathologic changes in the internal viscera The viscera of themselves have no sensation of pain as elicited by ordinary pain producing stimuli but when the viscera are irritated the stimuli are carried to the cord and react on the cord cells and impulses are produced and sent out as motor impulses or are carried to the brain by the neurons of these cells where they are produced as pain At the same time the adjacent set of cells become irritable and react abnormally to all stimuli reaching them from the periphery In this phenomenon lies the origin of the hyperalgesic zones of Head which are responsible for several peculiarities of pain production

Transferred pain may have varying degrees of two components (1) The stimulus passes from the neuron in which it is originally present over an intermediate neuron to a third neuron in the area of distribution of which it is perceived as being present (2) In other cases the sensorium mistakes the peripheral distribution of the pain as in degeneration of the posterior roots (tabes) or of the ganglia and posterior columns or cornu of the cord

Examples of pain transferred to a homologous segment in the same relative position on the opposite side of the abdomen are found in appendical and ovarian diseases pneumonia and pleurisy Examples of higher and lower reference are found in those cases in which the pain of pneumonia is transferred to the appendical region or in which the appendix causes pain which is transferred to the thorax

The term crises has been applied under certain conditions to paroxysmal attacks of pain of great severity It is accepted that a pain crisis consists of a paroxysm of pain as violent as human nature can endure accompanied by excessive functional activity of the part attacked but disappearing as rapidly as it appeared and that it is associated with a condition of undisturbed functional activity of the affected viscera between the paroxysms

Tissue susceptibility to pain is determined by the blood supply of the organs The organs which are the richest in blood supply generally suffer the greatest pain and with the exception of the bones the organs poor in blood supply have little if any pain

Clinical Evaluations of the Complaint of Pain Often much time is wasted if too long is spent on inquiries into details of location situation and distribution of pain the degree of itching and the occurrence of dizziness as complained of by the patient

Effeminate men and wordy women are inclined to exaggerate in recounting their symptoms A statement made with a cheerful countenance that the speaker is at the moment suffering horrible agony does not square with the facts the combination of cheerful manner and exaggerated speech is of diagnostic value as indicative of self deception hysteria or a habit of chronic overemphasis

In the majority of cases in which really severe pain is present certain symptoms are manifest that cannot be simulated The respiration is increased in rate the pupils are dilated the skin is wet with perspiration the pulse is likely to be tense there is a feeling of faintness and frequently a large amount of urine is passed within a brief period after onset

In the patients manner of describing pain there are differences that already have been implied Some patients as a matter of pride practice understatement of their subjective sensations whereas others from various motives habitually magnify their suffering and in most instances without the slightest intention of deception This mental attitude arises largely from a desire to obtain relief by impressing the physician with its urgent necessity

Variations in descriptions and perhaps in pain sensibility are racial as well as individual Persons of Semitic stock and members of Celtic and Italic groups appear

periods of time if it is of long total duration Freedom from pain is admitted without emotional difficulty by the patient and in the periods free from pain an attempt is made to return to accustomed activity (Walshe)

Histamine headaches are primarily due to dilatation and stretching of the pial and dural arteries and their surrounding tissues The large cerebral arteries at the base of the brain which include the internal carotid, the vertebral and the basilar arteries and the proximal segments of their main branches are chiefly responsible The fifth cranial nerves are the principal afferent pathways for the pain felt in the frontotemporal and parietal regions of the head The ninth and tenth cranial and the upper three cervical nerves are the afferent pathways for the pain felt in the occipital region of the head

In *migraine headache* aberrations in the tension of the extracranial and possibly the dural branches of external carotid arteries may be responsible for the cephalalgia It seems likely that the point of origin of the scotomas of migraine is situated in the cranial cavity

A *psychogenic headache* is commonly described as continuous and unremitting over long periods of weeks months or even years It responds to none of the usual analgesic drugs This headache is described by the psychoneurotic as terrible agonizing as if corkscrews knives or needles were piercing the head When questions as to locality and quality of the pain are pressed the patient s answers become vague and pains are sensations of weight pressure or constriction and not real pain at all In the hysteric these accounts are commonly given with animation and evident satisfaction while the anxiety neurotic is plunged in gloom and self pity In these patients there is no true pain but an image of pain conjured up by the patient and subjected to those processes of progressive elaboration to which mental images are prone

It has generally been believed that the majority of headaches *following spinal anesthesia* are the result of leakage of spinal fluid through the dural puncture with subsequent changes in cerebrospinal fluid dynamics loss of cushioning effect on the brain and resulting pain owing to pressure or traction, or both on sensitive brain structures and large vessels

The mechanism responsible for the production of headache after *lumbar puncture* is not clear At present there is no better theory than that accounting for the headache on the basis of a lowering of spinal fluid pressure by the removal of spinal fluid and loss of more fluid by leakage after the needle is withdrawn

Aberrations in the Central Projection of Pain *Propagated pains* which are felt in areas other than those in which they are produced are classified as associated referred projected reflex and transferred pains This classification is important to remember when the complaint is evaluated

Associated pain depends for its production on transference of stimuli from one nerve cell to another In some cases the means by which the stimuli are transferred cannot be determined

Referred pain is that class of pain in which the irritation occurs along the course of the nerve fibers and the pain is felt as being produced in the somatic peripheral distribution of the affected nerve or nerves

Sympathetic pain is really a transferred pain with the distinction that in sympathetic pain a painful sensation is present in the organ originating the pain whereas in transferred pain there may be no painful impression or sensation in the region or organ in which the pain originates

Projected pain is felt as being present either in a part that has no sensation (as in locomotor ataxia) or in a part which because of amputation no longer exists

Reflected (deflected or reflex) pain is that pain in which the stimulus is carried to the sensory ganglia or to the cord and then is transferred from the sensory filaments of the neuron primarily affected to those of a secondary neuron The stimulus

When a patient has a disorder induced by the physician through auto suggestion based on the physician s examination or manner of discussion of the disorder it is designated as an *iatrogenic disorder* A patient may have a disease or disorder induced by the technical procedures employed by physicians and specialists in examinations medications or operations these conditions are designated *iatrotechnical diseases or disorders*

Experience teaches that each topographic area of the body and each system of the body is liable to iatrogenic disorders or iatrotechnical disorders For instance locomotor disorders may be induced by the prolonged application of appliances even if properly fitted Improper fitting of appliances may be disastrous as illustrated by Volkmann s ischemic paralysis The use of unnecessary arch supports or the transfer of arch supports from one pair of shoes to a second pair of a different size or last is a common cause of complaint

The application of appliances for instance a stiff heavy back brace for a fatigued back without significant organic disease may originate a serious disability in a hypersensitive patient

A bunion operation in an occasional instance may leave a hypersensitive painful scar just as disabling as the original bunion and thus create an iatrotechnical disorder

Inquiries by a physician regarding the complaint of cough in association with expectoration hemoptysis or shortness of breath may initiate these symptoms for the first time in a hypersuggestible individual Certainly the continued treatment of an irritative cough in a hypersensitive individual may suggest to a patient a need for coughing Likewise the treatment for sinus troubles when there is only a postnasal drip fixes the disorder of sinus trouble in the patient s mind and the postnasal drip is complained of as sinus trouble

The recognition of the complaints indicative of hyperventilation and that of sighing respirations is more essential in many instances than the recognition of some such condition as a mild pulmonary emphysema If a patient has one of these syndromes it should be made clear to him that he has an insignificant illness resulting from a nervous tension and not disease of the lungs which prevents the entrance of a sufficient amount of air into the lungs

Perhaps there is no disease any better known and more dreaded by most intelligent patients than is pulmonary tuberculosis At times extensive study and observation may be required in order to determine whether or not the reinfection type of pulmonary tuberculosis is present If there be indeterminate symptoms present whether or not these are the result of an active pulmonary tuberculosis care is exercised lest the suggestible patient become overly concerned before an active pulmonary tuberculosis can be proved If there is bleeding from the nose or nasopharynx which is detectable in the sputum an ensuing cough often irritative and of a nervous origin may arise all of which may be prevented by diligence in explaining that the origin of the bleeding is the nose and not the lungs Even though great care is exercised by the physician in explaining an irritative neurogenic cough by the time the patient reaches home after having pondered the questions and the examinations made by the physician often an entirely erroneous conclusion is reached of what has been said The patient reasons that so many questions would not have been asked and so many examinations would not have been made just to explain an inconsequential cough There is no way for the physician to avoid these reactions in his patient except to recognize so far as possible the reactions of certain individuals and avoid any suggestive remarks concerning the organic bases for the complaints until sufficient objective data have been obtained or such data cannot be had

Syphilis in the past was more dreaded than tuberculosis and often uncertainties of diagnosis have created iatrogenic syphilophobia There are two sources of origin for iatrogenic syphilophobia (1) The possibility of having contracted syphilis car

to possess an average of greater sensibility to pain than those of Teutonic or Slavonic groups The important variations of course are personal or individual The neurotic patient of any racial group will complain bitterly of pain from a cause which should not give rise to more than simple discomfort The pain suffered by the abnormally sensitive person however is as real and existent in the consciousness as the slight discomfort felt from the same cause by a person given to less verbosity The pain suffered by an abnormally sensitive individual even though such pain in a normal subject might not be considered severe, is quite as disabling as a more severe pain in a more stoic individual

Manner of life and occupation probably do not modify susceptibility to pain The habitual endurance of hardship teaches the realities of life and trains the individual in the methods of proper evaluation of pain Conversely the person guarded from rude mental or physical contacts and not accustomed to discomfort and pain may feel that moderate pain is unbearable A strong mental prepossession such as religion or excitement over an accident may interfere temporarily with the registration of painful impressions on the consciousness Sensibility to pain is likely to be increased by its long continuance and it is a common observation that each recurrence of pain during the course of a disease finds the patient less able to bear it Fright or expectant apprehension invariably increases pain and sometimes originates it

Complaints Referable to Iatric Disorders The impression is often gained from eliciting the complaints of patients that they have got the terms definitions and expressions used in descriptions of their discomforts from observation or discussion with others who are sick or ailing In some instances they have acquired their terminologies from physicians

Information terminologies and misinterpretations gained from talking to relatives friends or other patients are not nearly so serious as those obtained from physicians or from others who administer to the sick and indigent When a patient goes to a physician this act is in itself an expression of confidence in that physician Whatever the physician tells the patient the patient believes in most instances Since the patient believes the physician this places a responsibility on the physician to mark his words when discussing a patient s disease or diagnosis with him

Honesty is moral soundness rectitude is that integrity chiefly applicable to social transactions sincerity is honesty of mind or intention The fact that patients are honest and sincere in their belief that they have physical complaints that can be rectified by injections treatments or surgical operations does not in itself justify the performance of such procedures *It is unfortunate that the carrying out of therapy by some* practitioners of the healing arts honest and sincere but not thoroughly grounded in the scientific principles of medicine and surgery has subjected patients unnecessarily to injections for toxic conditions and to surgical removal of appendices and gall bladders and in women of the internal genitalia

It is appropriate here to quote from Oliver W Holmes There is nothing men will not do there is nothing they have not done to recover their health and save their lives They have submitted to be half drowned in water and half choked with gases to be buried up to their chins in earth to be seared with hot irons like galley slaves to be crimped with knives like codfish to have needles thrust into their flesh and bonfires kindled on their skin to swallow all sorts of abominations and to pay for all this as if to be singed and scalded were a costly privilege as if blisters were a blessing and leeches a luxury What more can be asked to prove their honesty and sincerity? (*Medical Essays* Boston, pp 378 379)

Patients who have a heightened threshold for suggestion may become either alarmed or highly pleased at the suggestion of a particular diagnosis In either case what the physician has said about the diagnosis of a certain train of symptoms may create in the patient s mind a permanent disorder or an erroneous belief about the disease or disorder from which he suffers

may be told by the physician that his fatigue may indicate some organic disease Specialists in the diseases of the chest have emphasized the importance of fatigue in early pulmonary tuberculosis to the extent that the term fatigue is firmly associated with this disease However fatigue is common in low grade infections and more es pecially in the psychoneuroses

It is true that organic disease may be overlooked and the patient treated for functional disease but the reverse is more often true and frequently more disastrous

When surgical operation is indicated for organic disease in the presence of func tional disorders explain to the patients the difference between the two and do not leave the impression that repairing the lacerated perineum cauterizing the cervix suspending the uterus removing the gallbladder or the appendix is going to restore to them a stable nervous system The surgeon is too often pleased by the temporary relief due to the rest in bed following operation and by the diversion of patients minds from their troubles to their operations Unfortunately the relief thus obtained is not lasting

As has been stated iatrogenic and iatrotechnical disorders may occur in any topographic region or system of the body Physicians try to prevent them One help ful way in preventing them is to realize that the patient may be wrong in the report of what the foregoing physician has said or done

Concluding Remarks in Regard to the Complaint In an occasional instance it may be wise for the physician to disregard the initial complaint of the patient and to follow through chronologically each and every departure from health to the present time and then make his own conclusions how best to proceed

THE HISTORY ANAMNESIS

In the elaboration of the complaint much time as has been indicated in the foregoing discussion may be necessary in obtaining the *what* *when* *where* and *why* The *what* is the concept of the patient of the cause of the initiating factors of the illness the *when* is the time sequence of the events the *where* is the position of the patient in relation to place and person and whether or not the patient was affected inwardly or outwardly by the environment Environment here means the surround ing conditions influences or forces The classification of the what when and where constitutes the *why* and thus is the history

The history arbitrarily is divided into the present illness the past illnesses which may or may not pertain to the present illness and the family or genealogic history which likewise may or may not pertain to the present illness A classification of the attestations of the nature and purposes of the complaint may embrace all of the arbitrary divisions of the history but usually only the present illness

Before one begins the recording of the history it is well briefly to relate the complaints back to the patient and ask for corrections and elaborations If the events and complaints can be quickly verified by the patient the first definite piece of diag nostic information is at hand the complaints are based on organic disease If the patient heckles and makes many changes in and additions to the complaints when they are related back to him likely the disease or disorder is of functional or nervous origin Thenceforward the physician questions in his own mind the validity of all assertions and complaints the patient may utter

The History of the Present Illness The complaints having been obtained the present illness can be separated In most instances the present illness is but an elab oration in sequence of the origin and development of the complaint or complaints

During their training medical students commonly are admonished that in prac tice the physician must obtain a full and careful history in each case A full and careful history may not be obtained in some instances on one survey of the illness It is obtained or rather built up by repeated attempts often extending over days Emphasis may change when the patient has had time to ponder the questions asked

ries a severe social disapproval A physician who remarks to a self conscious patient that you may have syphilis may create a disorder more serious than syphilis itself (2) False positive serologic reactions The treatment of false positive serologic reac tions by antisyphilitic therapy often creates a serious iatrotechnical disorder (see False Positive Serologic Reactions Chapter 17)

Iatrogenic heart complaints and disorders may be found to have originated from the statement of some life insurance examiner or physician to the effect that the heart shows some abnormality such as a murmur or irregularity of rhythm as revealed by the electrocardiograph or else from the rejection of the applicant for life insurance on the score of some heart disturbance or of high blood pressure Sometimes it is a mere assumption on the part of the applicant himself that the heart must be diseased because two or three examiners were called in to listen to it In a person of the ap propriate mental composition the slightest suggestion that the heart is not intact may be enough to start a myriad of emotional reactions, which are very difficult to subdue if the patient has a relative or friend who happens to succumb suddenly to a *coronary occlusion*

An individual may have forgotten for a long time the remark of a doctor about some minor abnormality in the heart action until there appears some symptom which calls the attention of the patient to the heart and renews the doubt that might have once existed as to its integrity The renewed interest in the action of the heart may be created by a sudden skip or a twinge of pain or may be merely what is regarded as undue palpitation or dyspnea after some special exertion Such disturbing symp toms are often first noticed during convalescence from an illness

In eliciting the complaints of patients it is not to be supposed that organic heart disease is found only in individuals who have stable minds and nervous systems Organic heart disease does occur in those whose mental make up predisposes them to apprehensions and fears These patients will have a tendency to complaints ex plainable on the basis of organic heart disease and too there will be complaints and *symptoms which cannot reasonably be ascribed to the heart disease present* How ever when a structural disease is itself causing severe or distressing symptoms the neurotic symptoms if present at all are likely to be relatively unimportant It is in the milder forms of heart disease with few or no real symptoms that the complaints due to emotional reactions tend to occupy the foreground A correct differentiation between the complaints which are the legitimate result of the existing organic disease and those which are merely the expression of the associated psychic upheaval de mands skill judgment and experience Experience and common sense dictate just what is to be expected in the way of symptoms from the type and grade of the organic lesions present in order to recognize that certain of the symptoms have *no direct* relation to the somatic disease

This association of organic disease with complaints of a purely neurotic nature is encountered in cardiac patients of almost all ages Older children who have rheu matic heart disease have been taught by an overanxious mother or physician that all physical effort is to be avoided They complain of what they have been taught to expect if exercise is attempted Neurotic women who have long standing but rela tively mild mitral stenosis and persons in middle life who are oppressed by the knowledge that they suffer from high blood pressure may be full of complaints given to them by a physician the physician having diagnosed their heart disease by means of an electrocardiogram Physicians some of them are much too prone to regard physical overexertion as the most serious hazard to which cardiac patients are ex posed and to fail to give proper weight to the potentialities for harm that reside in *violent* psychic stimuli and prolonged sedentary habits

Iatrotechnical disorders and complaints often arise from the treatment of a pa tient for functional disorders as organic disease The patient is submitted to unneces sary medical and surgical treatment thereby intensifying the neurosis Such a patient

disease cannot be overestimated Each symptom should be elicited and the time of its appearance correlated with the chronology of the other symptoms

The statements of patients with reference to the therapeutic agents used by physicians previously consulted are ordinarily untrustworthy If such knowledge can be obtained from authoritative sources however it may be of considerable value Thus certain symptoms otherwise unaccounted for may be explained as due to the administration of certain drugs

The Past History The past history includes information about the patient the age sex nationality occupation places of residence habits experiences sensations and previous diseases or injuries Such considerations embrace also the chronologic occurrence seasonal or diurnal of certain diseases and the comparative infrequency of others These incidents are first entered in outline form and later translated chronologically as the history is written for permanent record

AGE Anatomic structure varies with age and physiologic processes have peculiarities which are characteristic of different periods of life Moreover the effects of environment occupation habits the beginning and end of sexual life and the wearing out of the organism are manifested at varying ages Consequently there is a distinct preponderance in the frequency of certain diseases or classes of disease at special age periods The diseases of youth are often the direct progenitors of those of old age and the life of the individual may be a constant struggle with disease of hereditary origin or with disease that began in prenatal life

SEX Putting aside the diseases due to differences in structure and function between men and women there remain certain maladies which have been said to occur more frequently in one sex than in the other These discrepancies are attributable mainly to manner of life and in women to disabilities incident to gestation Men have been said to suffer especially from diseases induced by exposure by hard physical or mental work and worry and by the acquirement of injurious habits Women more commonly than men lead an indoor life and many are harassed by household worries and domestic anxieties which create a special environment In the diagnosis of disease statements concerning differences in habits of men and women are of minimal value

RACE The susceptibility to disease or its opposite immunity possessed by certain races has caused much comment Often the data presented favoring racial immunity or susceptibility are not convincing for there have not always been clear definitions of the term race The terms race and nationality are not always synonymous (see Races Chapter 16)

OCCUPATION In considering the effects of occupation in causing disease it will be found necessary to ascertain the nature of the patient s employment whether active or sedentary and whether or not the work requires handling of injurious tools or handling of toxic or irritating substances or inhaling them Hobbies habits and contacts with animals are details a knowledge of which may be useful Previous occupations should be ascertained The state of health sometimes enforces the occupation

RESIDENCE Knowledge of the place of the patient s residence may be of considerable importance if not with regard to diagnosis at least with regard to prophylaxis of future attacks In the diagnosis of the fevers the fact of the patient having visited or lived in countries or localities where these diseases are prevalent may furnish a clue otherwise lacking Malaria hookworm fungi and rickettsial disease are examples of diseases which have at times special affinities with certain localities

Other items embraced under the head of residence concern the effects of climate of residence in city or country seashore or inland and the sanitary condition of dwellings with reference to ventilation drainage heating cleanliness water and food supply and the availability of sleeping nets for protection from insects

HABITS The habits formed by individual persons are interwoven with age occu

Likewise the recorder of the history will desire to inquire about other happenings after time has passed He tries first of all to get all the complaints arranged according to the systems from which they originate in generalized disease he attributes them to the body as a whole emphasis then will usually be obvious

The historic data having been arranged according to topography or to the system of origin the etiology if not already evident is sought In obtaining data pertaining to etiology direct interrogation is necessary It is mandatory to reduce the number of etiologic possibilities to a minimum In making this reduction it is well to realize that the etiology of all diseases has been categorized in the various nomenclatures of diseases into (1) diseases due to prenatal influences (2) infections (3) intoxica tions (4) trauma or physical agents (5) diseases secondary to circulatory disturb ances (6) disturbances of innervation or psychic control (7) disturbances of static mechanical abnormality (8) disorders of metabolism growth or nutrition (9) new growths and (10) those diseases in which structural reaction is manifest but the cause is unknown Some hereditary and familial diseases belong in the last category

It would be repetitious to give here in greater detail than the foregoing outline the reasons for placing any particular disease or condition of topography or system in a certain etiologic category for that is the object of the text of this book

It is well to realize that in obtaining the history the largest drafts are made on the tact of the physician and it is during this time that the important relationships be tween physician and patient are being established The physician is not a prose lytizer the right of the patient to his own religion and morals is granted Success depends to a great degree on true friendliness on mutual confidence and essentially on correct mutual understanding of the spoken word

The patient may be one of the odd persons from whom it is difficult to extract more than a monosyllabic answer or he may be so talkative that a question can be slipped in by the physician only when the patient pauses to breathe Dense ignorance may be an obstacle so also may shame or false modesty Exaggeration of symptoms a too common failing must be quickly recognized and likewise the opposite a stoic pride in making light of symptoms and disability These and other difficulties for example malingering require the exercise of skill in the art of cross-examination and diplomacy

Except in the case of suspected malingering when the answer may flatly contra dict the alleged condition leading questions are to be avoided especially with im pressionable or ignorant patients For instance it is better to say Did you have any pain in the head? than You had pain in the head did you not? The first question is a simple interrogation which may elicit a reply of Yes or No The second almost forces the answer Yes Care is to be taken lest the patient s story should be too narrowly limited otherwise a knowledge of important symptoms may not be gained It is well to expend additional time and patience and if required to lower questions to basic or even to depraved linguistic levels in order to make sure that a possible vital point in the history is not missed

An important point with reference to early diagnosis of a host of diseases of infectious origin is to learn of any known exposure Another important diagnostic aid is information about fatigue of mind or body dietetic imprudences toxic agents and chilling of the body or taking cold This last factor is frequently assigned by patients as a satisfactory etiologic explanation of the most diverse ailments

A definite statement of the time of onset will generally place the disease in one of two categories acute or chronic As a rule acute diseases begin suddenly whereas chronic maladies may continue over a long period before the symptoms force them selves on the attention of the patient

The manner of onset should be accurately ascertained by separating the symp toms which initiated the attack from those which appeared at a later period The value of a strict chronologic history of the symptoms which succeed the onset of any

ing diseases in their prognosis or as suggestive in regard to their possible occurrence in a given case deficiency states myocarditis cirrhosis of the liver chronic pharyngitis chronic gastritis delirium tremens neuritis and Korsakoff s disease

The Family History The family history of the patient is of much importance because of the light which may be cast by it not only on the present illness but also on the constitution and tendencies of the patient

It is not always possible to obtain a complete and accurate family history It is usually necessary to cross-examine the patient with some particularity inquiring into the symptoms and duration of illnesses attributed to ancestors and bearing in mind the approximate meanings of various common terms For instance old age is frequently assigned as a cause of death from the age of 50 years or more and is a term which has little meaning In many instances in which childbirth is assigned as a lethal cause death is found to be due to some other cause

Inquiries regarding certain diseases should be made cautiously because of the possibility of arousing feelings of shame or fear If it is apparent that shame and fear exist it is better to ask about the symptoms without mentioning the names of suspected diseases such as syphilis tuberculosis and cancer A feeling of reproach to family or personal pride in acknowledgment of the existence of certain ailments may lead the patient to conceal important information

A full statement of the family history includes the nature of the illnesses (with the age of the living) and the causes of deaths (with the age at death) which may have occurred among the patient s parents paternal and maternal grandparents brothers and sisters It is requisite at times to ascertain similar facts with reference to aunts uncles and cousins It should be borne in mind that transmissible tendencies may pass over one generation and may be sex linked Also it is just as important to know and to inform a young couple whose first child is born defective that malformations are often sporadic In an isolated case of a congenitally defective child in a known normal genealogic history there is no reason to suspect that subsequent children will be abnormal Occasionally however contributing factors can be discovered if the basic information is at hand In general the study of the causes of congenital malformation has yielded definite knowledge concerning some causative factors It may be pointed out that German measles (rubella) toxoplasmosis and irradiation are known causes of congenital malformations

COMMENTS

While discussing the complaint and the history the physician forms a conception of the mental characteristics of the patient in health and as modified by disease As a rule the more highly trained the mind by private study or at college or in business and the more elaborate and complex the social environment the greater is the necessity for an appreciation of the personal traits of the individual Influences which produce no apparent effect on some laborers toughened by hard life will cause profound perturbation in the somewhat hyperesthetic person man or woman whose nervous system is in a state of tension from the demands of family society art literature or religion

If necessary information may be obtained from friends or relatives concerning the mental traits of the patient in health but such information should not be accepted without proper consideration of its source Inquire especially as to the existence of extreme susceptibility to pain and of exaggeration in the manner of expression as to whether the patient is or is not easily elated or depressed or given to undue solicitude about personal health of himself and of others and other points which may be serviceable in estimating the value of the statements made by the patient or the present deviation from the normal mental and physical health Such inquiries should be made with circumspection and tact in order to avoid misconception of motives

pation and residence The possible existence of addiction to alcohol, opium cocaine or other drugs is always present Diet the use of tea coffee and tobacco clothing sleep and exercise are largely governed by the social condition and environment Both men and women should be interrogated as to the amount and strength of tea and coffee taken daily The tea and cake habit is mainly found among women The examiner should ascertain also the kind and amount of alcoholic beverages taken and the time of taking that is, whether before during or between meals and how much tobacco is used of what kind and in what manner (with reference to naso pharyngeal catarrh nervousness and cardiac neuroses)

The experiences of an individual depend on many factors among which are per sonality type and physical stamina The experiences and sensations are recorded only if pertinent to the present illness

PREVIOUS DISEASES OR INJURIES A knowledge of prior illnesses and injuries is of value provided that their date nature and severity can be ascertained for three reasons

1 An attack of any of certain diseases renders subsequent attacks of that disease probable Among such diseases are angina pectoris apoplexy appendicitis asthma pneumonia bronchitis tonsillitis colics (gallbladder and renal) convulsions (in fantile or epileptic) malaria gout migraine and neuralgias

2 With many of the epidemic fevers of bacterial viral and rickettsial origin a previous attack may negative its subsequent occurrence

3 A history of the previous existence of certain diseases or injuries may throw light on present conditions which stand in the relation of sequelae to the primary ailments Examples of these diseases are syphilis gonorrhea scarlet fever rheuma tism and chorea and septic or suppurating foci leading to subsequent embolic or general inflammations of heart lungs liver pleura bones or peritoneum any or all of them

A history of a fall or other injury may be of some value in arriving at a diagnosis for instance of a protruded intervertebral disk jacksonian epilepsy or sympathetic ophthalmia A previous surgical operation may point to the possibility of a recur rence of the condition which required operative intervention

There is little of diagnostic value to be gained from the varying statistics of the *seasonal prevalence of disease* in the United States beyond the broad statement that diarrheal diseases predominate during the summer months and poliomyelitis in late summer whereas pulmonary disorders and rheumatic affections are most prevalent in the winter and early spring Zymotic diseases occur in largest number during the cold season but this is to be explained rather by the opening of the schools and the closing of house windows than by the effect of season in itself

It may be mentioned here that certain diseases either begin or show an *exacer bation of symptoms at special diurnal periods* Bronchial asthma is likely to make its onset or to intensify in severity in the early morning hours spasmodic croup as well as diphtheritic stenosis of the larynx between 10 P M and 12 P M The suffer ing from painful diseases is usually worse at night and in febrile disorders the tem perature generally reaches its highest point between 7 P M and 8 P M The paroxysms of whooping cough are more frequent and severe at night than at other periods The pain due to diseases of the bones joints and peripheral nerves presents a nocturnal aggravation Pain of rheumatoid arthritis is likely to be worse when the patient arises in the morning than at other times

It is well to remember that some ailments present a more or less regular *periodi city of recurrence* Among these are neuralgias migraine epilepsy periodic paralysis relapsing fever malarial infections ulcerative endocarditis paroxysmal hemoglobin uria pyelonephritis bronchial asthma and menstrual disorders

A history of *alcoholism* is important as explanatory of the presence of the follow

ing diseases in their prognosis or is suggestive in regard to their possible occurrence in a given case: deficiency states, myocarditis, cirrhosis of the liver, chronic pharyngitis, chronic gastritis, delirium tremens, neuritis and Korsakoff's disease.

The Family History. *The family history of the patient is of much importance* because of the light which may be cast by it not only on the present illness but also on the constitution and tendencies of the patient.

It is not always possible to obtain a complete and accurate family history. It is usually necessary to cross-examine the patient with some particularity, inquiring into the symptoms and duration of illnesses attributed to ancestors and bearing in mind the approximate meanings of various common terms. For instance, old age is frequently assigned as a cause of death from the age of 50 years or more and is a term which has little meaning. In many instances in which childbirth is assigned as a lethal cause, death is found to be due to some other cause.

Inquiries regarding certain diseases should be made cautiously because of the possibility of arousing feelings of shame or fear. If it is apparent that shame and fear exist, it is better to ask about the symptoms [illegible] the names of suspected diseases such as syphilis, tuberculosis and cancer. [illegible] *of reproach to* family or personal pride in acknowledgment of [illegible] *may*

[illegible]

and the charge of making imputations uncomplimentary to the patient The physician in private practice has favorable opportunity to please and help because in consulting him the patient has exercised choice in seeking personal advice The physician in institutional practice conducted by a group or a governmental agency has more difficulty because the patient may not have had a choice of physician

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3

EXAMINATION

For the general examination the patient is fully undressed of the usual clothing An appropriate dress for the examination is a cape about the shoulders and a sheet around the waist Many other suitable garments have been devised and all are satisfactory

Manner and Dress The physician begins to observe the manner and dress of each of his patients as soon as acquaintance is made

The manner of a patient pertains to the method of performing or executing the ordinary social acts over which custom presides Each individual has a peculiar mode of performing these acts depending on the training which has been molded by the environment to which he is accustomed This definition of manner assumes that the individual under observation is reasonably free from physical discomforts and mental stresses The psychic and physical stresses of illness often change the manners of both men and women For instance an individual who is suffering from hysteria under normal conditions of life may under the adversities of an additional illness to himself or to a member of his family behave in a stable and admirable manner An individual of good manners when in health may express a bad manner under the stress of illness and pain in himself or in members of his family

Modern dress is no longer an expression of status or determinate function There remains a well developed sense of decorating the body with clothing and accessories such as jewelry and furs In individual instances it is frequently difficult to differentiate between clothing and ornament this is especially true of the headwear of women Cultists with distinctive mental warping may dress foolishly solely for the purpose of exhibition justifying such dress by reference to the Scriptures as If I may touch but his clothes I shall be whole (*Mark V*)

Although clothing no longer is a symbol of class distinction it may have diagnostic implication in the individual or in groups of individuals The most significant implication is related to the sexual representations or aspirations of the female During the last 200 years women s dress has changed constantly and alternately in an endeavor to conceal or to reveal the back breasts shoulders arms legs and thighs During the period of a particular vogue all women good bad and indifferent will *and should conform to the fashion The physician is not interested in* fashion or in those who conform to fashion but in those who do not conform Of definite interest to the diagnostician are two small groups namely those who become conspicuous by showing more of the thighs than fashion permits and those non-conformists who show none of the thighs despite fashion Those who show too many charms are likely to be involved in sexual and marital intrigue or think they might like to be These patients often bear the burdens of moral and social diseases as well as being maladjusted sexually It must be realized that many physically beautiful and delicately and tastefully dressed women are sexually cold Those who show none of their charms are sociologically maladjusted and may be affected by all sorts of abnormal ergasias

Of the accessory articles of dress, tinted spectacles when worn because of weak eyes if there is no disease of the eyes indicate that the wearer admits poor health Excessive and gaudy jewelry even expensive ornaments of excellent workmanship

often adorns selfish persons Perfumes in profusion except among Latins may indicate the absence of the regular use of soap Distinctive highly aromatic perfumes are often used by those afflicted with homosexual and heterosexual practices

In the evaluation of patients with regard to their clothes the examiner has in mind that with the majority the influence of social customs prevails as expressed in the Arabian proverb Eat what you may but dress as others do

An occasional hint may be derived from care of the dress Omission to use fastenings which may be needed for common decency the coat or trousers buttoned with the wrong buttons or a vest soiled with droppings of food may indicate the mental enfeeblement due to psychosis or chronic alcoholism or due to the physical disability resulting from arthritis or paralysis Clothing wet and of ammoniacal odor is observed in cases of incontinence of urine and cystitis when urine dries it may leave a white deposit on the clothing A patient may be so crowded and bulging in his clothes as to suggest a recent rapid increase of bulk from obesity or general dropsy The shoes may be left partly or entirely unfastened from forgetfulness gout rheumatism or edema or slit for similar reasons or because of corns bunions or injury or worn more on one side or in front or at the heel because of paralysis deformity or disease of the joints

A great deal may be gleaned from the behavior of an individual when asked to disrobe for the examination A proper amount of timidity at having to undress for an examination is expected of both sexes Extremes of timidity may interfere with or prevent a satisfactory examination If the examination is rendered unsatisfactory or impossible this should be made clear to the patient Insistence and persistence never accomplish anything Patience on the part of the examiner with full explanation of the reason for an examination usually gains the desired results

During the examination the examiner extends his knowledge and attempts to confirm those things which have been learned from the complaint and the history He employs his own functions of knowing combined with his feelings and his strivings to learn and to demonstrate wherein the trouble lies He looks (inspection) he feels (palpation), he listens (auscultation) to determine the condition of the tissues These primary methods of sight touch and hearing have many applications and combinations the simplest of which is percussion a combination of touch and hearing Sight touch and hearing are the primary methods of examination of the patient It should be realized that native capacity for employing the senses varies with each person Only a few have the natural capacity of Laennec Trousseau and Flint and it is these few who do not argue about the relative merits of palpation auscultation and percussion versus roentgenologic examination or any other way of objectively demonstrating derangement of tissue by trauma or disease These few physicians and surgeons who have natural ability do not argue they are happy to find for themselves the source of disease or they cheerfully accept the help that anyone or any appliance or other means can give them Any person endowed with an average degree of the sense perceptions and intelligence can learn the technic of studying and examining patients A lack of acuity of one of these senses may be overcome by constant effort practice and development of the other senses For all it must be known that at least three out of four illnesses are diagnosed and treated from the diagnostic data obtained by the physician from the use of his senses and his knowledge of disease

Theory and Practice of Inspection Inspection which is begun with the first sight of the patient is the most valuable method of examination During the taking of the history the sex approximate age race posture facial expression nutrition color of skin eyes and hair and many other details are observed During any part of the taking of a history and the performance of an examination the examiner requests Let me see you do it or Show me where the pain originates and where it goes

The light preferably daylight when feasible, as a rule should fall directly on the surface to be examined Changes in direction from which the light comes controlled by moving either the patient or the source of the light so as to permit the rays to fall obliquely across the surface to be examined are often of value by production of shadows and may accord emphasis of much use in detecting small pulsations or abnormalities of shape and movement

Theory and Practice of Palpation Palpation the use of the tactile sense of the hand and fingers is an indispensable means of obtaining information in all manners and means of diagnosis The remarkable extent to which the educated touch may be developed is exemplified in many physicians

In the practice of palpation care should be taken to keep the nails short The hands should always be warmed before touching the patient All rough or abrupt pressure is avoided The hand at first should be laid flat on the part later the fingers and their tips may be employed to determine and localize more accurately any discoverable abnormality When palpating deep seated organs or when there is a large amount of overlying fat or fluid the palpating hand may be usefully reinforced by pressing on it with the other thus sparing the palpating hand much muscular exertion and leaving unimpaired its power of tactile appreciation

When palpating the points to be determined with reference to the part or organ are temperature shape size measured (stated in metric terms) consistency movability with or not with respiration tenderness superficial or deep seated fluctuation the undulation of an enclosed fluid produced by the pressure of the fingers and appreciated by another finger placed on the opposite side of the enclosing cavity fremitus pulsations thrills or vibrations originating in the heart or blood vessels

The synchronous use of the methods of inspection and palpation is to be encouraged When these are used separately always look before feeling—and feel before listening

Theory and Practice of Auscultation Auscultation may be performed by direct application of the ear to the body or it may be aided by use of the stethoscope Auscultation may be combined with inspection and palpation (see Diseases of the Respiratory System Chapter 9)

Directions and Topography In the examination of patients directional terms for description of findings are employed constantly and it therefore is important to use the proper terms that do not permit confusion Such terms as *front* and *back* *top* and *bottom* are not desirable For the trunk *dorsal* and *ventral* *cranial* and *caudal* are the proper directional terms but are not generally employed *anterior* and *posterior* *upper* and *lower* *apex* and *base* are the terms in use though they may admit of confusion *Dorsal* and *ventral* are permissible but not wholly admirable terms applicable to the inferior extremity and the upper arm as well for these aspects are constant in orientation with the trunk The terms *superior* and *inferior* are so firmly entrenched in diagnostic descriptions particularly in connection with aspects of the vertebrae and as names of structures (as M serratus posterior superior) that they cannot be abandoned The term *superior extremity* is diagnostic and almost always is used instead of the more correct designation *pectoral extremity* The meaning of *anterior* and *posterior* when applied to aspects of the ankle is likewise clear but when applied to the wrist it is not clear for the wrist is reversed in pronation and supination In designating aspects of the wrist *dorsal* and *volar* (or *palmar*) are the proper terms

Cranial and *caudal* though technically proper terms for the aspects of the head are clumsy The head of man is oriented on the same axis as that of quadrupeds and *anterior* and *posterior* *superior* and *inferior* are permissible terms *Lateral* and *medial* directions in the upper arm and in the inferior extremity may be used although *fibular* and *tibial* in referring to sides of the lower leg and the foot are prefer

able *Lateral* and *medial* directions are objectionable for forearm and hand because it is not known whether the natural or the anatomic positions are meant For these *radial* and *ulnar* directions are correct

Directional terms of a general nature in antithetic pairs are as follows (based on Howell s *Gross Anatomy* D Appleton Century Co 1939)

Anterior and *Posterior* front and back respectively properly used for the head only but permissible at times because of established usage for the trunk The proper corresponding terms except for the head are the following

Dorsal and *Ventral* The term dorsal indicates the back the entire back or upper side of an animal when the ventral surface is down Dorsal is incorrectly used to designate a particular segment of the back for instance that of the thorax Likewise ventral is not for the front of the thorax The thoracic vertebrae are often incorrectly designated dorsal vertebrae Since vertebrae are identified according to the region of the back to which they belong for instance as cervical thoracic lumbar sacral and coccygeal it is incorrect to designate any of them as dorsal vertebrae They are thoracic vertebrae In instances of increases over the usual number of vertebrae in a particular region the extra vertebra is designated by addition to the usual number for that region for instance a sixth lumbar vertebra The dorsal aspect of the limbs both upper and lower is the part covered by the dorsal or extensor musculature The dorsal aspect of the arm is the back of the upper arm back of hand and wrist and the corresponding aspect of the forearm The dorsal aspect of the inferior extremity is the front of the thigh leg and upper part of the foot

Dorsal and *Volar* (or *Palmar*) the proper terms for back and palm respectively of hand and corresponding aspects of forearm

Dorsal and *Plantar* the proper terms for upper aspect and sole respectively of the foot

Superior and *Inferior* permissible but less desirable (except for head) alternatives for cranial and caudal

Cranial and *Caudal* the proper terms for the indicated directions in the trunk

Superficial and *Deep* superficial is nearer the surface (from any aspect) as opposed to deep

Medial and *Lateral* medial is toward the median plane and lateral is toward the side

Peripheral and *Central* peripheral toward the periphery central away from the periphery Used mostly in connection with the nerves

Proximal and *Distal* proximal toward the median plane distal away from the center Used mostly in connection with the extremities and the alimentary tract In the alimentary tract proximal is employed to designate nearest to the mouth or to the beginning of the segment irrespective of topographic situation For instance *proximal jejunum* indicates that part nearest to the stomach or the mouth even in the presence of situs inversus viscerum In the alimentary tract particularly the intestines *distal* indicates away from a designated point It is used in opposition to proximal Common use of these terms is in connection with artificial stomas for instance proximal stoma the one which discharges the intestinal dejecta *Distal stoma* designates the direction in which the intestinal contents would or do flow away from the stoma

External and *Internal* used chiefly to indicate respective surfaces of hollow organs or of the body as a whole

Radial and *Ulnar* the proper terms for denoting the sides of forearm wrist and hand toward the radius and toward the ulna respectively

Tibial and *Fibular* the proper terms for denoting the sides of the lower leg ankle and foot toward the tibia or toward the fibula respectively

The following terms in frequent use might prove confusing unless explained

Median in the median plane

Mediad toward the median plane

Medius middle, between dorsal and ventral or between external and internal

Intermedius between medial and lateral

Transversus transverse to a particular part of the body

Transversalis transverse to the body axis

Transversarius relating to some structure which is transverse (as Mm inter transversarii the muscles between transverse processes)

As the body is three dimensional so there are three planes of the body

Sagittal Plane the dorsoventral longitudinal plane through or parallel to the sagittal suture

Coronal or *Frontal Plane* the longitudinal plane from side to side at a right angle to the sagittal suture through or parallel to the coronal suture

Transverse Plane the horizontal plane at any level at right angles to both sagittal and coronal planes

Anatomic Regions The primary regions of the body as discussed here, comprise *head* (caput) *neck* (collum) *back* and *superior* and *inferior extremities* Each of these is customarily divided into secondary surface regions The boundaries of some of these are not always to be determined with accuracy by one inexperienced in anatomy for the reason that their extent usually depends on some subsurface feature Anyone interested will find by reference to the *Basle Anatomical Nomenclature* (B N A) list of regions that many of the secondary regions (as parietal frontal deltoid posterior femoral and the like) are self apparent and that some other terms are of but little use in diagnosis

In this text each of these regions is described in association with the topography and topographic diagnosis when that particular region is being considered Generally the requirements of the *Basle Anatomical Nomenclature* are at least respected and partially fulfilled

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4

TOPOGRAPHY AND REGIONAL DIAGNOSIS OF DISEASES OF THE HEAD AND ORGANS OF SPECIAL SENSES

The general practitioner and the internist are not expected to make specialized examinations and diagnoses or to carry on specialized treatment However they are required to have sufficient knowledge of diseases described and diagnosed by the specialist to discuss intelligently with the patient the diagnosis and treatment which the specialist suggests in the individual case In the discussion that follows it is hoped that there is information which may be of some help to the physician in his fulfillment of his obligations to the patient

The head varies in shape and size according to race sex and age As a rule the head of a man is larger than that of a woman The head tends to change shape and increase in size with age If the head enlarges out of proportion to the rest of the body the condition is designated as *macrocephalia* If the enlargement tends to produce mainly a broad head the condition is designated as *brachycephalia* If the enlargement tends to produce a conical shaped head the condition may be designated as *oxycephalia* If the head is unusually long from front to back the condition is known as *dolichocephalism* If the head is unusually narrow the condition is designated as *stenocephalia* if the head is unusually small as *microcephalia* and if the head is generally distorted and of abnormal shape it may be designated as *plagiocephalism*

In many men past middle age an increasing prominence of the supra orbital ridges develops with the result that the eyebrows seem to overhang the eyes more and more and the countenance looks different from what it did 10 or 20 years earlier This enlargement is due to or is associated with enlargement of the air cells of the frontal sinuses and is not pathologic However if this enlargement of the forehead is excessive it must be distinguished from acromegalia and from another condition of no clinical significance formerly designated leontiasis Leontiasis may or may not be associated with a thickening of the frontal bone which is known as benign frontal exostosis this too is of no clinical significance Often a patient who has enlargement of the forehead seeks medical advice with the complaint that he has noticed that the size of his face is increasing such a patient nowadays wonders whether or not he has disease of the pituitary gland The condition can be distinguished from acromegalia in that rarely in acromegalia is the frontal bone alone affected and more often the affection of the forehead is much slighter than the increase in the size of the lower jaw and the phalanges of the hands and feet Tumors of the frontal bones almost always cause an asymmetric enlargement

Changes of Shape and Contour of the Head in Disease In certain diseases characteristic changes in the size and contour of the head and face are of diagnostic importance

In *hydrocephalus* the head is abnormally large and usually globular sometimes pyramidal in shape Hydrocephalus is most frequently observed in infants and children An abnormal increase in the measurement of the head is the principal symptom by which chronic hydrocephalus can be recognized Aside from the en

largement the most characteristic feature is the prominence of the forehead at the root of the nose The rachitic head may resemble a hydrocephalic head Many children recover from mild degrees of hydrocephalus The only evidence of it left in adulthood is an increase in the size of the forehead which gives the appearance of a rather large dome shaped head

Heads deformed by *rickets* and changes of the head produced by *sporadic cretinism* or *myxedema* are described in appropriate places in subsequent chapters

If the face presents an appearance as if it were composed of lateral halves from two different individuals with the vertical line of junction sharply defined and if the hair of the smaller side is thin or absent the eye sunken and the skin altered in appearance it is probably an instance of facial *hemiatrophy*

If both sides of the face appear normal but one is much the larger the condition known as *congenital hemihypertrophy* should be suspected Along with congenital hypertrophy hemangioma of the skin over the affected regions often is present Since congenital hemihypertrophy often involves one half of the body confirmation of congenital hemihypertrophy should be sought by making comparative measurements of the arms and legs The arm and the leg which are hypertrophied may be on the same side as the hypertrophied half of the face or they may be on the opposite side

Facial asymmetry may exist also with congenital torticollis or wryneck If the cranium is enlarged and globular and the malar prominences are marked and the orbital rims massive the presence of hyperostosis of the cranium should be suspected

The tuberous growth of *leprosy* occurring on the face together with the resulting ulceration and cicatrization slowly changes the shape and contour of the countenance which assumes the so called leonine aspect or facies leonina

Tower shaped skulls are rather characteristic of some of the congenital anemias (see Hemic Diseases Chapter 11)

Abnormal Movements of the Head *Nodding spasm* is a rhythmic nodding movement of the head In children this movement may be a form of habit spasm or of epilepsy In cases of epilepsy or convulsive disorders nodding spasm is accompanied by a momentary loss of consciousness It may occur in hysteria either alone or as a part of the so called *hysterical chorea*

Pulsating shaking of the head which occasionally is seen in the aged is not necessarily evidence of senility or arteriosclerosis It seems at times to have a hereditary factor It is often found in postencephalitis or Parkinson s syndrome

Spasmodic torticollis is a spasmodic jerking of the head which recurs every few minutes In the chronic form of the disorder the head is brought toward one shoulder which may be simultaneously jerked upward to meet the head at the same time that the face is rotated to the opposite side and the chin is raised In chorea as contrasted with spasmodic torticollis the movements of the head are extremely irregular and bizarre and the facial muscles are affected before those of the neck Torticollis is often a form of conversion hysteria

Inability or disinclination to move the head especially if the patient is prone to support it by hand may be due to disease of the cervical vertebrae If this manifestation is associated with dysphagia it points to disease of the articulation between the atlas and the occiput

Retraction of the head of a bedridden patient—that is a drawing back and *fixation* of the head by contraction of the posterior cervical muscles so that the occiput bores into the pillow—is a symptom in all forms of cerebral and cerebrospinal meningitis particularly when the disease is affecting the basal meninges It is seen in tetanus and in strychnine poisoning as a part of the general tonic spasm

Other causes of abnormal fixity of the head are occipital cervical myalgia and cervical arthritis swollen and painful cervical lymph nodes sprain of the cervical muscles or other traumatism of the neck postpharyngeal abscesses congenital and

spasmodic torticollis and contracted cicatrices especially extensive scarring from burns

THE SCALP

The *skin* of the scalp is the thickest in the body although not so dense as that of the heel Besides the hair follicles the scalp contains abundant sweat and sebaceous glands and blood vessels which are associated with the hair follicles

The *hair* of the scalp is to a very slight extent an index to the general robustness and vigor of the individual Thick coarse hair is at times associated with strength of constitution and scanty fine textured hair with a delicate habit of body but there are many instances in which this is not the case There is some diagnostic value in an observation of the color and scantiness of the hair (see page 1197)

Loss of Hair Color Graying of the hair the physiologic loss of the pigment of the hair begins in the average individual at 40 to 45 years of age and slowly increases as the years pass Early grayness is not always a sign of premature old age and is not necessarily associated with degenerative arterial changes Gray hair is compatible with good health and otherwise normal tissues The loss of color is frequently dependent on a hereditary factor

When chronic poisoning is suspected if it is apparent that the hair is dyed—and dyeing usually is apparent—it may be an important clue in the diagnosis (see Hair Dyeing Chapter 19)

Alopecia (Acomia Calvities Baldness) The etiology of alopecia is not often obvious Congenital baldness (alopecia adnata) is rare This is usually associated with defective teeth and nails These two conditions are of little practical importance diagnostically

Premature Alopecia This is an exceedingly common condition especially in men It is endocrine in origin eunuchs are never bald It is not attributable to modern civilization or to the wearing of tight hats It is familial in origin and common to all races of men

Premature alopecia begins at about the age of 20 years and is often completed by the age of 30 years only a rim of hair being left at the back and sides Cases however do occur in which the loss of hair is more gradual and even on the vertex may never become complete The scalp may be obviously seborrheic or covered with dandruff which undoubtedly aggravates the condition or it may look normal

A corresponding type of alopecia occurs in women but has several points of difference from that seen in men It is more general and is not limited to the vertex is not so nearly complete and is more regularly accompanied by seborrhea of the scalp

Alopecia Without Scarring The etiology in alopecia without scarring is unknown A disturbance in innervation or psychic control is possible In susceptible persons a shock may precipitate an attack during World War II attacks occurred in three or four weeks after the patients had been torpedoed or bombed Reflex irritation from neuralgia a carious tooth septic sore throat or similar condition has been blamed but these are not valid causes The patients even those who have alopecia totalis are usually in excellent health

There are several varieties of alopecia without scarring for instance alopecia areata trichorrhexis nodosa and trichotillomania

Alopecia Areata This common condition is characterized by circular areas of baldness It is not related to the color of the hair and it affects men and women about equally In Anderson s experience there was a difference in the position of the primary patch between the sexes in men it was 60 per cent occipital 25 per cent frontovertical in women it was 27 per cent occipital 56 per cent frontovertical A positive family history was present in 19 per cent but it is doubtful that this indicates

a familial tendency Other members of the family were affected at the same time in 6 per cent of cases Vitiligo was present in 4 per cent of cases

The affected hairs show thinning toward the roots and are easily removed with brush and comb They may also break off a few millimeters above the skin surface

Alopecia areata is a relapsing disease beginning in childhood but more severe between 20 and 29 years of age The hair slowly returns to the bald spots though it may be delayed for months and may grow back white and remain white When the disease steadily progresses until all hair all over the body is lost, the alopecia usually is permanent Another resistant form of the disease is that in which the scalp margin is involved in a patchy fashion These patches may coalesce to form a band, giving a wiglike appearance to the rest of the hair

Nail changes are common in extensive alopecia When the nails are affected the changes are usually in the form of rows of tiny pits as is sometimes observed in psoriasis These pits often are unnoticed by the patient and are found only on routine examination Nails of both the hands and the feet may be affected

Alopecia areata is differentiated from ringworm by the absence of scaliness of the scalp and of fungus in the hairs Cicatricial alopecia offers no difficulty Furunculosis is eliminated by the fact that the scar of the original lesion has a center of a red dot

Prognosis is good in ordinary cases for patients less than 40 years of age When there is involvement away from the scalp for example of eyebrows and the bearded areas and around the edges of the scalp the lesions are stubborn and persistent Recovery is slow also in cases in which scattered white hairs are present on the bald patches Chronicity is universally a bad omen Alopecia totalis is to be feared in all cases in which the disease lasts more than 18 months

Toxic Alopecia There are many toxic agents including roentgen rays and radium that cause a fall of hair After prolonged fevers the hair may fall Falling hair results from a similar cause during the secondary stage of syphilis (moth eaten scalp) and after pregnancy After gestation the onset may be sudden and the fall severe In all toxic alopecias an important diagnostic point is that the hair behind the ears can easily be pulled out

Trichorrhexis Nodosa This condition is due to too frequent use of too strong soaps and excessive friction as may result from a habit of rubbing the hair The hairs show white nodes which on microscopic examination are seen to consist of a splitting of the hair structure so that it has the appearance of having the end worn off

Trichotillomania Trichotillomania is a habit comparable to nail biting Hairs are consciously or unconsciously pulled out The irritation of pediculosis may start the habit An area or areas of normal scalp dotted with normal hairs of varying length and a surprising amount of alopecia are the result The diagnosis requires roentgenoscopic examination of the stomach in search for the possible presence of trichobezoar (hairball)

Alopecia With Scarring The causes of alopecia with scarring comprise all the etiologic agents producing scarring such as roentgen rays radium burns and infections such as favus kerion of ringworm or lupus erythematosus The primary cause is usually recognizable The atrophy following the use of roentgen rays or radium is a factitial dermatitis the scars of burns are deep and contracted favus has a long history of gradual spread and may still show marginal activity kerion scars are circular or are combinations of circles lupus erythematosus may show activity or scarring over the bridge of the nose and medial sides of the cheeks

Tumors The scalp contains an abundant blood supply and consequently *telangiectasis* and venous and cirsoid aneurysms are frequent (see page 316)

There are a number of tumors of the scalp of congenital origin Cephalocele a protrusion of part of the cranial contents may be of congenital or of acquired origin

The cephalocele occurs in the occipital and sincipital regions either above or below the tentorium An *encephalocele* is a hernia of brain matter and is covered with the arachnoid it protrudes through a congenital or traumatic opening in the skull A *meningocele* is a cystic mass a protrusion of the meninges covered by pia mater A *hydrencephalocele* is a protrusion of a lateral ventricle or of the fourth ventricle covered by brain tissue and the arachnoid

Dermoid tumors which consist of occluded ectoderm occur in the median line of the head and face They often appear over the anterior fontanel and the external occipital protuberance Often a thin pedicle attaches the tumor to the dura mater

Cysts Sebaceous epidermoid inclusion dermoid and mucous cysts occur about the head and neck

Cystic skin tumors whose contents consist of keratin are keratomas and those which are of acquired origin and which contain sebaceous material are sebaceous cysts Inclusion dermoids are congenital cysts developing along the embryonic lines of fusion and containing hair follicles sweat glands and sebaceous glands in their walls Epidermoids are similar in nature to dermoid cysts with the exception that their linings possess no dermal structures

The wens or sebaceous cysts usually occur in the scalp and in the lobe and on the posterior surface of the ear The common tumor of the adult scalp is the sebaceous cyst which sometimes becomes calcified Such tumors are usually multiple and are rounded firm and though freely movable are invariably attached to the skin at one point a feature which often distinguishes them from dermoid and epidermoid cysts In the skin overlying many keratomas and sebaceous cysts there is visible an enlarged pore from which cheesy material can be expressed on manipulation of the growth There is a wide degree of variability in the dimensions of these tumors some are scarcely palpable nodules while others attain a diameter of several centimeters

Keratomas and sebaceous cysts ordinarily symptomless are frequently the site of an acute inflammatory process in which the tumor becomes excessively tender and painful and objectively manifests a sudden increase in size When acutely infected the cyst resembles an acute abscess and exhibits all of the general and local symptoms which attend such a process After an acute infection subsides a keratoma or sebaceous cyst may resume its normal proportions and appearance In an occasional case the infection completely destroys the epithelial lining and with subsequent healing the entire cavity of the growth is obliterated Often an acute infection in one of these tumors becomes chronic with the result that purulent material is continually discharged externally through a sinus producing excoriation of the surrounding skin Occasionally one of these cysts becomes chronically infected and then is surrounded by dense fibrocicatricial tissue which may completely mask the true nature of the lesion

THE FACE

The face embraces the anterior part of the head The bony framework of the face is made up of some of the bones of the skull as well as those of the face proper Deformities and disfigurements of the face which result from injuries or disease or both assume a greater importance to the victims than comparable troubles would elsewhere for many persons feel that their face is their fortune

The right and left halves of the face differ from each other and present varying degrees of asymmetry This facial asymmetry seems to be present in all races In the right half of the face the features are said to be more characteristic of the individual and are more conspicuous than in the left The right eye is more prominent than the left eye a fact which may be related to right handedness In the left half of the face the individual features recede

Malformations The relatively frequent occurrence of malformations of the face is explained by the complexity of the developmental processes in this region

nerve supply to a portion of the skin Eating or drinking is accompanied by excessive perspiration in the area in which the nerve supply has been impaired Langenskiold offered what he believed to be a satisfactory explanation of this distressing symptom the preganglionic sympathetic fibers governing sweating had been destroyed thus according to Cannon s law the postganglionic cholinergic fibers were rendered hyperexcitable and during mastication acetycholine elaborated in the parotid gland diffundated to the nerve endings in the skin and produced sweating Gustatory asymmetric hyperhidrosis may occur below the neck as described by Mellinkoff and Mellinkoff

THE MUSCLES OF THE FACE

Facial Spasms Spasmodic contraction of the facial muscles may be tonic that is continuous or clonic that is intermittent It may be unilateral or bilateral and may involve one or all of the muscles which are innervated by the facial nerve In order to form an opinion of the cause of spasmodic affections of the face it is advisable to take into consideration the age and sex of the patient and to ascertain the presence or absence of disease of the eyes teeth and skin In some cases the facial symptoms constitute a significant part of a disease of the central nervous system

The diseases or conditions which may be indicated by facial spasm are mimic spasm convulsive tic blepharospasm nictitating spasm exophthalmic goiter chorea epilepsy tetanus meningitis hysteria spasm following paralysis irritation at the root of the facial nerve and finally lesions of the cortical center of the facial nerve

Mimic spasm is usually bilateral and ordinarily does not begin until adult age A more or less constant twitching of one side of the face with partial closing of the eye occurring in an adult is a mimic spasm

Habit spasm usually occurs in neurotic children Characteristically during the spasm there is a sudden wink of the eye or a rapid drawing of the mouth to one side or a sniff occurring at irregular intervals and greatly intensified in severity by emotional causes

Convulsive tic ordinarily is a hereditary affair occurring in neurotic children The tic is irregular movement of the facial muscles and frequently those of the arms and sometimes is accompanied with an explosive ejaculatory utterance of profane or obscene words or this tic may consist of involuntary repetition of a word or the mimicking of an action

FACIAL PAIN

Infections of the sinuses and the antrums cause facial pain Dental neuritis particularly when it occurs in senile patients is troublesome and unfortunately is not readily relieved Often improvement of the general health of the patient by appropriate dietary regimens ameliorates the complaint The *localized dental neuritis* that follows procaine block injections is due to the excessive amount of procaine injected into the immediate vicinity of the nerve The symptoms of paresthesia usually disappear Postherpetic *ophthalmic neuralgia* is persistent and its course is not affected by operative procedures Snake venom may help

The neuralgias are common causes of pain in the face In *glossopharyngeal neuralgia* the pains are projected along the distribution of the ninth cranial nerve The trigger zone is situated in the tonsillar fossa and the darting pains are projected toward the ear usually terminating in the tympanum The pains are precipitated by yawning swallowing and talking but not by rubbing the face The pains resemble those of trigeminal neuralgia in their occurrence duration repetition and character but they differ from those of trigeminal neuralgia in their distribution In glossopharyngeal neuralgia it will be observed that cocainization of the pharynx will result

in temporary cessation of all attacks but cocainization of the pharynx will not alter the paroxysms when they are due to trigeminal neuralgia

Sphenopalatine neuralgia is characterized by boring pain situated in the superior maxillary region It is most pronounced under the malar bone and at times extends into the gums of the superior maxilla the hard palate and downward into the neck This pain differs from that of trigeminal neuralgia in that it is more or less constant it is not paroxysmal and is moderately severe but it is not sufficiently severe to incapacitate the patient unless there is an accompanying neurosis Neither deep injection of alcohol into the branches of the trigeminal nerve nor division of the sensory root affords relief in the treatment of this type of neuralgia

The trigeminus nerve (trifacial) arises in the medulla oblongata along the floor of the fourth ventricle and supplies motor fibers to the masseters pterygoid and the temporal muscles and muscles of the tongue It furnishes the sensory fibers to the skin of the face (Fig 4 1) and to the tongue teeth and the cornea The three divisions of the nerve are designated the ophthalmic the maxillary and the mandibular

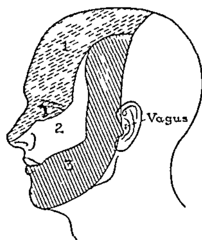


Fig 4 1 Areas within which sensory disturbances and pain may occur in diseases of the trigeminal (trifacial) or fifth cranial nerve 1 ophthalmic division 2 maxillary division 3 mandibular division

Local lesions of the nerve rarely cause severe pain However local lesions are often accompanied with impairment of sensation within the sensory distribution of the nerve for instance in the mouth loss of corneal reflex or numbness of the skin

Trigeminal neuralgia (tic douloureux) is of unknown etiology and usually occurs in persons more than 50 years of age It may occur in an occasional person after the age of 40 years

The chief symptoms are the intense unilateral darting pains which radiate from the side of the nose or the upper lip through the teeth or into the eye or even into the forehead and head The pains come in paroxysms lasting for a few minutes during which the face is flushed and the eyes and nose run There is an expression of agony The patient once the disease has started is seldom free from some pain and a breath of cold air speaking eating or putting out the tongue may bring on a paroxysm The pains may come on for a certain length of time each year often during the beginning of cold weather

On examination there may be observed spasmodic movements of the face or jaws There is no disturbance in the functions of the trigeminal nerve in either the sensory or the motor spheres At the sites where the divisions of the nerves pass through the fascial sheaths on the upper lip and in the mouth may be observed a so called trigger point On stimulation of a trigger point pain immediately ensues The employment of ice for the precipitation of pain in one of these patients is a cruel practice

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The infection as it occurs in children is not a serious one There are moderate fever and mild congestion of the nose and throat which precede the swelling of the parotid glands The pain in mumps is produced by increased glandular activity This fact accounts for the time honored procedure of a test for mumps favored by grandmothers for many generations of having a child who is suspected of having mumps bite a pickle After the age of puberty mumps is a more serious disease especially in boys and men because of the frequent and painful complication of orchitis which may lead to testicular atrophy and to sterility Oophoritis is less serious because the inflamed and edematous ovaries are not confined by a thick fibrous tunica like the testicles in adolescent boys or in men There may be a minimal involvement of the parotid glands the first symptoms being those of orchitis which in mumps may be either unilateral or bilateral The orchitis often is accompanied with acute severe pain and there may be a high fever In women a mild mastitis may occur

Examination There may be either a unilateral or a bilateral enlargement of the parotid or submaxillary glands or none discernible on examination Swelling of a gland usually reaches a maximum within 48 hours after onset and most often persists for 7 to 10 days Occasionally some enlargement of the glands of salivation can be discerned after the lapse of several weeks

In the earlier stages a restricted area of redness may be observed immediately about the orifices of the ducts of Stensen and Wharton Fever of moderate degree may be present 12 to 24 hours before swelling is observed and it usually persists from 1 to 3 days In many cases fever is absent

The most important findings on examination in mumps are those indicating involvement of the epididymis and testicle This may be bilateral or unilateral and when bilateral sterility often ensues There are exquisite tenderness pain and swelling of the testicles If the testes are involved on subsidence of the swelling atrophy may be observed either unilaterally or bilaterally On the basis of studies on changes in the blood diastase and amylase some degree of pancreatic involvement is often revealed Physical findings may reveal evidence of encephalitis during the course of mumps (see Diseases of the Brain Chapter 15)

The total leukocyte count may be moderately elevated or depressed but is usually within normal limits The differential count may reveal an absolute or a relative increase in lymphocytes from the first to the fourteenth day of the illness In contrast to the blood the leukocyte count of the cerebrospinal fluid is of great significance in the diagnosis of the general syndrome of aseptic lymphocytic meningo encephalitis of which mumps virus is one of the known etiologic agents In mumps encephalitis the number of white cells of the fluid is increased The total count may range from normal to more than 2 000 per cubic millimeter In a majority of cases the proportion of lymphocytes is from 90 to 100 per cent It is known however that in apparently uncomplicated encephalitis a moderate or marked increase in lymphocytes may occur in the spinal fluid A specific complement fixing antibody may be present in the blood of adults and children who deny having had mumps

Diagnosis Often the parents or the grandmother has diagnosed mumps correctly before the physician is consulted if he is consulted at all The diagnosis in sporadic cases is difficult for it is necessary to differentiate mumps from suppurative parotitis enlargements due to foreign bodies in the salivary ducts neoplasms Mikulicz s disease and uveoparotid fever The possible complications are numerous and appropriate diagnostic examinations and procedures will depend on the existing complication

It is now possible to obtain serologic evidence of any type of infection caused by mumps virus This can be accomplished by demonstrating the appearance of or increase in an antibody capable of fixing complement in the presence of mumps

antigen and by the demonstration of the appearance or increase of a specific factor antihemagglutinin in the blood serum which inhibits the agglutination of chicken or human erythrocytes by mumps antigen The first procedure is recommended

As with many serologic tests, a definite diagnosis can be made only as the disease progresses and there is a significant rise in antibody titer, revealed by successive examinations of the blood following the onset of the disease

Chronic Inflammation Chronic inflammation of the parotid gland may remain symptomless When symptoms do occur there is a sudden swelling and pain of the gland which commences during a meal The swelling and pain subsides in 3 to 4 hours

On examination during the attack the gland is greatly enlarged and tender and no secretion is discharged from the duct Between attacks the secretion is thick and glairy Subsequent attacks may occur frequently they do not The glands may be enlarged

Tumors Mixed Tumors Mixed tumors of the parotid gland are hard usually ovoid and lobulated and are situated in front of the ear and below it Tumors of this type invariably develop in the lower half of the gland whereas adenomas and primary malignant growths of the parotid gland usually originate in the upper half of the gland In their early stages mixed tumors are situated within the gland substance and consequently are not freely movable The adenomas are freely movable until they become malignant

Mixed tumors of the parotid gland develop in a congenital anomaly and seldom are discovered before early adult life Usually such a tumor is not seen by the physician until years have elapsed and the patient has known of its presence for a long time As the tumor enlarges it causes a rounded induration in the parotid region Situated as it is within the substance of the gland the tumor in the course of growth sometimes causes a stretch on the facial nerve so that partial paralysis of the nerve takes place Complete facial paralysis as the result of a parotid tumor is unusual however even if malignant changes have occurred within the tumor If the mass attains a large size the skin over it may become ulcerated The ulceration causes very little pain A mixed tumor of the parotid gland grows slowly its size is not an indication of whether or not it is malignant and palpation will not determine this point A search for affected lymph nodes in the region therefore is important

Mixed tumors that lie superficially to the parotid gland tend to form pendant growths in other words tumors thus situated grow away from the gland rather than within it

Adenomas and Cysts Adenomas are fairly common in the parotid gland where they begin as simple encapsulated nodules of gland tissue As stated they tend to occur in the middle and upper parts of the gland seldom in the lower pole These cystic glandlike tumors become malignant much more frequently than do the mixed tumors of the parotid gland Prior to the malignant change they grow slowly

Carcinoma Carcinoma of the parotid gland often begins within the normal gland tissue or in adenomas If a simple parotid adenoma has been incompletely removed surgically the remaining adenomatous tissue seems to be a favored site for malignant disease and a carcinoma occurring in such tissue has a bad prognosis Carcinomas in adenomatous rests or within the gland substance become extensive regions of infiltration involving much of the gland and the tissues beyond it Carcinomatous tumors of the parotid gland are hard irregular masses but they cannot be diagnosed by palpation They are diagnosed by histologic study of biopsy material The prognosis in malignant disease of the parotid is always serious

THE LIPS

Thick lips or thin lips are often a racial characteristic as in the Negro (thick) or the American Indian (thin) (see Races Chapter 16) Tradition has it that full

lips in white races mark a phlegmatic temperament The lips are thickened and coarse in myxedema and cretinism

The color of the lips is often significant of anemia and other conditions in which the skin is pallid and the first evidence of cyanosis is often seen in them their slight blueness attracting attention to the existence of cardiac or pulmonary disease

Open and particularly *dry and cyanosed lips* are likely to indicate dyspnea Open lips may be due to disease within the mouth stomatitis glossitis cancrum oris phlegmonous tonsillitis or some form of nasal stenosis Loose and pendulous lips in a very ill patient are suggestive of a central nervous system disorder that is early hemiplegia or bulbar palsy The open mouth is seen in some normal persons in all conditions of great prostration and in idiots and some insane persons

Trembling or twitching of the lips may be a symptom of general nerve fatigue paralysis or chronic bulbar palsy Convulsive raising of the upper lip with an accompanying squinting of the eyes and wrinkling of the forehead is occasionally evidence of great pain

The term *macrocheilia* implies thickened lips The condition is observed in children and usually the increase in size of the lips is due to an increase in the muscle and connective tissue In this condition the lips seem to be less prone than the tongue gradually to increase in size When the lips have attained two or three times their normal size the growth ceases and the size remains stationary The increase in size may be so little in evidence that the disorder is neglected until the child reaches a self-conscious age This great increase in the size of the lips occasionally accompanied by increased size of the tongue and the cheek is very much like the increases in size seen in hemihypertrophy which may involve a whole side of the body

Unilateral deviation of the lips the angle of the mouth being drawn to one side and downward if not due to loss of teeth on the opposite side or to the contraction of scars is indicative of facial paralysis either existing alone or as a part of a hemiplegia

A brawny hot *swelling of the lip* may be a small abscess or a more extensive and serious carbuncular inflammation The lips are swollen as a part of the disease in the fortunately rare cancrum oris Swelling may be due to the taking of corrosive poisons in which case the interior of the mouth will also be swollen and reddened Bites of insects may explain a swelled lip and among other causes are alveolar abscess bitten lip in epileptic or other convulsive disorder and angioneurotic edema

Foam on the lips is a common symptom during an epileptic seizure and sometimes in the later stages of cerebral apoplexy and in pulmonary edema It should be borne in mind that a bit of soap in the mouth is employed by malingerers in simulating epileptic convulsions

Malformations The zone indicating the meeting of the median nasal processes during development is normally evident by double ridges and a shallow groove passing downward from the nasal septum to the margin of the upper lip the *philtrum*

A median defect of the upper lip or jaw through incomplete union of the median nasal processes is a rare anomaly An equally rare anomaly is imperfect fusion at the midplane between the mandibular processes which form the lower lips and jaw A slanting cleft may extend from the mouth up the cheek It is due to the failure of union between a maxillary process and the nasal processes

Cleft Lip or Harelip This is a common disfiguring congenital deformity of the face It is so named because of its resemblance to the lip of a hare It is frequently associated with a cleft of the soft palate or the hard palate or both The deformity *is the result of a failure of union* of the embryologic components that after union differentiate the nasal and buccal cavities The first or mandibular arch very early in embryonic life becomes separated into a short upper maxillary process and a

longer lower mandibular process The maxillary process together with that of the other side and the intervening portion known as the premaxillary (frontonasal) process which has descended in the median line from the head unite and form the superior and lateral boundaries of the oral cavity and the nose If the premaxillary process unites with only one maxillary process a unilateral cleft is found but if it unites with neither a bilateral cleft is present the premaxillary process extends forward as a proboscis and a cleft exists in the alveolar ridge It can thus be seen that all gradations of a deformity may exist from a slight V in the lip to a complete cleft of lip alveolar ridge and palate The main symptom of harelip and cleft palate is nasal voice On examination the degree of disfigurement is proportionate to the degree of the congenital deformity The diagnosis is obvious

Inflammations Inflammations of either the upper lip or the lower lip may be the result of any ordinary pyogenic infection which has entered the lip through an abrasion The *staphylococcus* is a common offender in these inflammations

Inflammation of the lower lip is not nearly so important clinically as inflammation of the upper lip since a localized furuncle or a diffuse inflammation of the upper lip may extend and produce a sinus thrombosis with subsequent death of the patient Any inflammation of the upper lip of any magnitude at all should be considered as serious These lesions are much more serious in their import than are the malignant tumors of the lip which have received much more attention from members of the medical profession

Herpes (Virus Infections) The herpes virus infections of man occur in a variety of clinical types Herpes simplex groups of viruses may cause (1) diseases of the skin (2) diseases of mucous membranes (3) diseases of the eyes (4) diseases of the nervous system and (5) herpes progenitalis

One of the commonest lesions of the lip is *herpes febrilis* The etiology of herpes febrilis the common herpetic vesicles or coldsores or fever blisters is a filtrable virus that can be demonstrated by inoculating the corneas of rabbits The virus of herpes febrilis (herpes simplex) has been thought to be identical with that causing encephalitis lethargica and consequently it has been studied intensively

The patient who has a recurrent attack of fever blisters is as a rule singularly free from severe systemic symptoms It seems that such persons have become carriers of the virus and are likely after nonspecific stimuli such as fever menstruation or emotional upsets to have recurrent manifestations of herpetic infection

The lesion of herpes simplex is heralded by a feeling of irritation or burning pain in the area of skin involved Erythema quickly followed by papulation and vesiculation ensues in this area The vesicles are grouped thin walled lesions which rupture soon in from a few hours to a day The lesions may occur anywhere on the skin but definitely tend to appear most commonly at mucocutaneous junctions on the lips and on buccal surfaces of the mouth and to recur in the same sites The lesions heal without scar formation

Fissures Fissures of the lip sometimes follow trauma Less often a fissure remains after the healing of herpes labialis Generally fissures of the lip result after prolonged drying of the mucosa as a consequence of being out of doors in the heat and sun and dust most of the time or inside where there is considerable drying of the lips from exposure to high temperatures and low humidity Fissures may remain as such or they may become ulcerated

On examination of a chronic fissure it is seen that the fissure has formed and has healed again and again without there being any manifestation of malignancy However although a fissure may remain for years without notable change malignant degeneration may begin at any time along its borders Unless a fissure is picked with the finger nails it usually does not bleed When a fissure begins to bleed rather easily it is time to consider that a cancer may be arising in the cleft

Fissure of the lip may become painful the patient complaining of the pain for example after taking a chew of tobacco or a dip of snuff when the tobacco laden saliva gets into the fissure

Perleche Perleche as the term is now employed usually denotes the changes at the angles of the mouth which occur in vitamin B complex deficiency However idiopathic perleche and pseudoperleche do occur

Idiopathic perlèche includes instances of perleche encountered devoid of con comitant clinical or laboratory findings which in the light of present knowledge must simply be considered as of idiopathic origin In some instances there is an ac companying salivary drooling or possibly excessive or chemically altered salivary secretion in association with scrotal tongue with geographic tongue or with macro glossia

Pseudoperleche includes those instances of contact dermatitis neurodermatitis seborrheic dermatitis atopic dermatitis severe chapping and herpes which occur in the region of the oral commissures and because of the movement in this area by mastication and talking perleche like fissuring ensues Perleche like involvement is also seen at times in the rhagades of congenital syphilis and as an extension to this area from mucous patches of syphilis in the mouth Transitorily perleche like in volvement may result from the trauma of manipulations incidental to dental therapy

The symptoms of perleche are mild usually consisting merely of a feeling of dryness and at times a slight burning sensation Deep infected fissures may be pain ful and though rarely encountered occur chiefly in perleche of adults

Objectively the epithelium of one or both labial commissures is somewhat mac- erated either adherent or easily detached and wrinkled Later the wrinkles become deeper often forming one or more transverse fissures with little if any tendency to bleed The involvement usually stops rather abruptly just within the mucocutaneous junction of the commissure but extends as a localized erythematous scaling derma titis usually with fissuring from a few millimeters to as much as 2 cm or more out ward and downward from the mouth angle onto the skin of the lower lip Untreated the lesions in all types of perleche have a tendency toward spontaneous remission and exacerbation but they rarely disappear completely

Pyogenic Ulcers Pyogenic ulcers of the lips are a not uncommon end result of various traumas to the lips Some of these ulcers result from direct trauma such as a blow to the lip from a fist or being thrown against the windshield in an auto- mobile accident On examination the elevated surface is soft to touch and the whole lesion is soft when it is grasped between the fingers If the lesion is of long duration it may be hard on palpation Very little bleeding occurs from manipulation and *handling of these lesions and the adjacent tissues are but slightly hyperemic*

Some of these ulcers may be tuberculous in origin Tuberculosis of the lip occurs on the vermilion border without a like lesion elsewhere in the mouth Biopsy may be required for diagnosis

Benign Ulcers Benign ulcers occur either on the upper lip or on the lower lip On examination these spontaneous ulcers usually are manifested only by a defect in the epithelium Such lesions may continue for months even years without notice able change or they may heal only to recur Recurrent lesions may end in malig nant disease Ulcers of this type sometimes occur on the lower lip but not usually as the result of irritation from a pipestem Biopsy may be required for diagnosis

Chancres The primary lesion of syphilis occurring as a chancre on the lip, on the tongue in the mouth or on the tonsil is rarely seen in private practice Such lesions do occur and they should be watched for but it is erroneous to consider every ulcer on the lip in the mouth or on the tonsil as a chancre until it has been proved other wise A chancre may occur on either the upper or the lower lip When chancres of the lip do occur they frequently are on the upper lip Malignant lesions on the con

trary commonly occur on the lower lip Chancres develop and may attain considerable size within a few weeks Carcinomas of the lip do not progress so rapidly

On examination a chancre of the lip if it has been present for some time, has an indurated feeling to the examining finger and it is elevated and may be sharply circumscribed If a chancre is examined in its early stages, it is a small nonelevated lesion with but little induration about its edges

Positive serologic reactions of the blood or the spinal fluid conjoined with an ulcer of the lip in a young person, may be presumptive evidence that the ulcer is a chancre Positive serologic reactions with an ulcer on the lower lip in an older person may not be of diagnostic significance

Lesions which are the result of undernutrition may appear on the lip as punched out ulcers and it is likely that similar lesions will be found on the tongue or elsewhere in the mouth

Wounds Wounds of the lips when properly approximated heal readily because of the free blood supply This free blood supply also accounts for the extensive swelling which occurs after injury or infection the latter however only rarely occurring after injury

Tumors The commonest tumors of the lips are the benign cysts that form in the glands of the mucosa These are small bluish tumors occurring near the vermilion border more frequently of the lower lip On examination the dome of the benign cyst is firm and uniform in outline and it may or may not be translucent

Lymphangiomas Some of these lesions are congenital in origin but may not be manifested during the first several years of life They may occupy a part or all of the lip Although soft to touch they are not reduced in size by pressure If a lymphangioma results from trauma as is often the case in middle aged persons there is a firm region which is due to the scar which resulted from the trauma There is little difficulty encountered in the diagnosis of the lymphangiomas of congenital origin for they are uniformly boggy and compressible to the touch without being reducible in size Some lymphangiomas of the lip attain enormous size and are grossly disfiguring

Angiomas In infants the blood vessels mainly the veins of the lips sometimes become enlarged forming a large protrusion The initial enlargement may be noticed at birth or soon after birth as a dusky blue slightly swollen spot on the lip As the child grows the swelling enlarges sometimes rapidly

Hemangiomas These lesions are of congenital origin and are always reddish or purple On palpation they are soft and compressible and are reduced in size or obliterated by pressure In children they may be increased in size when the patient cries or coughs also in adults who have extremely large hemangiomas on the lips coughing increases the size of the tumors

Cutaneous Horns Cutaneous horns of the lips are labial lesions common in advancing years they may occur however in young adult persons Cutaneous horns of the lips do not differ from cutaneous horns on any other part of the body except that on the lips they are more likely to become malignant than elsewhere Over the rest of the body the horns may occur in successive generations and be shed

The first appearance of a cutaneous horn is a small elevation hard to the touch with a sharply defined base Occasionally a cutaneous horn will develop on the base of an ulcer or in a fissure or a granuloma These horns vary considerably in consistency one part may be very hard and cornified and another part may be granulomatous Some of the very small horns may be moderately hard They grow in length rather than in diameter consequently their name When inflammation exists around the base of these tumors it should excite suspicion that the tumor is malignant It is remarkable how long a patient will put up with one of these long excrescences on the lip In generalized cutaneous horns there is no known treatment Roentgen therapy is not effective

Pigmented Moles Occasionally along the mucocutaneous border of either the upper lip or the lower lip there will be a darkly pigmented mole Under no circumstances should a specimen of such a lesion be taken for biopsy These moles are potentially malignant Some of them are highly malignant (melano-epitheliomas)

Cancer Cancer or epithelioma of the lip almost always affects the lower lip rather than the upper lip It is commonly *secondary to a chronic fissure leukoplakia* or *keratosis* of the lip and it is a disease usually of middle aged farmers who are exposed to sun wind and drying of the lips These lesions are hard indurated ulcerating or bleeding and often produce considerable stiffening of the surrounding tissues

The disease extends through the lymphatic channels which pass down and out from the lips to the submaxillary lymph nodes and then to the nodes along the great vessels of the neck It is in these regions that the lymph nodes are affected by the spread of cancer from the lip The middle of the lower lip drains into a node in the submental region in front of the submaxillary nodes Finding this node enlarged is not of much significance for it frequently is enlarged when there is no demonstrable lesion of the lip Enlarged nodes may be found on both sides of the neck and are secondary to cancer of the lip

A *primary cancer of the lip* is defined as one developing without a previously existing fissure or ulcer It occurs more frequently in men than in women and about 10 times as frequently on the lower as on the upper lip Cancer of the lower lip is almost invariably of squamous cell type On the upper lip it frequently represents invasion of the lip by a carcinoma of the skin and may be either squamous cell or basal cell carcinoma

Primary cancer of the lip is a localized hard deep-seated lesion of firm density and is unyielding on pressure Examination of the epithelium over the lesion reveals that the surface is being approached or that there already is ulceration When ulceration of these lesions occurs there is free bleeding on the slightest trauma palpation of these hard ulcers causes them to bleed A patient may be unaware of the seriousness of an ulcer on the lip until the ulcer begins to pain or to become deeply ulcerated or the lymph nodes under the jaw become ulcerated

Liljencrantz classifies cancer of the lip by stages in its development determined on examination clinical stage I lesion less than 1 cm in diameter without muscle infiltration stage II moderate sized lesion (1 to 3 cm in diameter) usually accompanied by early muscle infiltration stage III large lesion (more than 3 cm in diameter) with deep infiltration and all lesions with involvement of bone or cervical lymph nodes

Cervical lymph nodes may contain cancer cells and not be enlarged Ulcerated infected primary lesions often cause inflammatory hyperplasia and it is frequently impossible to distinguish clinically between inflammatory and metastatic cervical nodes

Biopsy establishes the diagnosis Any ulcer fissure scaling heaped up or papillomatous lesion remaining on the lip for more than one month should be suspected of potential malignancy Inspection and gentle palpation are most important in making a clinical diagnosis A negative blood serologic reaction and absence of history rule out coexistent syphilis A positive serologic reaction does not exclude cancer and should not be the cause for delaying biopsy

THE MOUTH

Surface Anatomy In looking into the mouth one sees the tongue below and the roof above surrounded in front and on the sides by the teeth On each side are the inner surfaces of the cheeks and posteriorly are seen the uvula the arches of the palate and the pharynx On the mucous membrane of each cheek opposite the second upper molar tooth is a small papilla through the top of which opens the duct of the parotid gland A small probe can be inserted into the duct from the oral cavity and passed outward and backward toward the gland

Size of the Mouth The primitive slit may fail to reduce normally (macrostomia) on the contrary the degree of closure may be excessive (microstomia) Size of the mouth also seems to be hereditary

The Buccal Cavity Odor of the Breath (*Halitosis*) There are many variations from the normal in the odor of the breath Local conditions in the mouth may be accountable for an unpleasant or foul odor of the breath In dehydration when sordes have collected on the teeth the breath is likely to be stale and musty A foul odor attends in some degree all forms of stomatitis and glossitis and is most fetid in the mercurial and gangrenous forms Caries of the teeth necrosis of the jaw pharyngeal or tonsillar diphtheria follicular tonsillitis and lacunar concretions are local conditions that may explain the existence of unpleasant emanations Frequently a bad breath attends migraine headaches and chronic alcoholism A hot feverish breath is common in febrile disorders An ammoniacal urinous odor is not uncommon in the severer grades of uremia A heavy sweetish odor is perceived in bad cases of diabetes mellitus often preceding or accompanying diabetic coma

It is well to recall however that the normal human breath is odorless and that many instances of halitosis are the so called essential halitosis It has been said that man s breath becomes infected by the bad quality of food by the bad state of the teeth and still more by old age At one time or another during life everyone suffers from malodors coming from the mouth The presence of offensive breath does not cause pain or other subjective inconvenience If the patient is aware of halitosis, it may be the cause of deep despondency which arises because in most instances the foul odor is not at all recognized or at most only partially so by the patient

There is a popular opinion abroad that bad breath originates most frequently from disorders of the stomach and colon—constipation Gas cannot pass from the stomach or from the intestine back through the collapsed esophagus under normal conditions except in belching In certain diseases of the esophagus in which there is retention of food in a dilated portion or in a diverticulum there may be foul odors on the breath Pyloric stenosis and the late stages of cancerous ulceration of the stomach are productive of pronounced foul odors In cases of fecal vomiting and gastrocolic fistulas a fecal odor of the expired air may be observed

Dryness of the Mouth Dry mouth due to diminished secretion of saliva may lead to redness soreness and fissuring of the mucous membranes and if long continued eventually to atrophy In severe instances rapid deterioration of the teeth follows Chewing and swallowing may become difficult Inadequate amounts of saliva according to Allington may result from (1) a decrease in salivary gland substance (congenital hypoplasia senile atrophy destruction following surgical intervention irradiation inflammation or neoplastic infiltration) (2) disturbances of innervation (central or peripheral nerve lesions emotional states psychoses interference with conduction of nerve impulses due to drugs toxic agents or metabolic disturbances) (3) dehydration and (4) unknown causes (Sjogren s syndrome) In *Sjogren s syndrome* dry mouth is associated with dryness of the eyes and of the mucous membranes of the nose throat and vagina and with deficient gastric secretion Many of these features suggest deficiencies of vitamin A nicotinic acid and riboflavin and some of them are seen in pernicious anemia iron deficiency anemia and *Plummer Vinson syndrome* There is a frequent association of the syndrome with rheumatoid arthritis and with the endocrine changes that occur in postmenopausal women In aging men after many years of excessive smoking chewing of tobacco or dipping snuff dry mouth is common Treatment of dry mouth must be directed toward the causative factor

Salivary Taste Hormones Dryness of the mouth from any cause often impairs taste *Reasons for this clinical observation are partially accounted for by dryness of the mouth in some and in others by the hereditary absence of salivary taste hormones* It seems that most persons can taste saccharin and sodium chlor e on a

wet tongue but some cannot taste these substances on a dry tongue Cohen and Ogdon have demonstrated a salivary taste hormone which could account for these discrepancies of taste on dry tongues and in some a normal absence of this hormone

The Sense of Taste The four primary taste sensations are bitter sweet acid and salt

Taste buds the end organs of the gustatory sense cannot be identified grossly They are scattered over the mucous membrane of the mouth and tongue at irregular intervals and occur especially in the sides of the vallate papillae They decrease in number gradually during adult life and rapidly in extremely old age

The taste fibrils run in both the glossopharyngeal nerve and the trigeminus nerve the former supplying the posterior third of the tongue the latter the anterior two thirds A complete loss of taste may result from trigeminal disease alone and occasionally from lesions affecting only the glossopharyngeal nerve It seems therefore that the gustatory fibers may at times be present solely in the trigeminus and at other times solely in the glossopharyngeal nerve Some of the taste fibers may pass by way of the chorda tympani since a partial loss of taste may be present in facial paralysis Ordinarily a solution of sugar is sufficient to enable one to detect any impairment of taste

The disorders of the gustatory sense are ageusia (loss or impairment of the sense of taste) and parageusia (perversion of the sense of taste)

AGEUSIA Impairment or absence of the sense of taste may be due to local un healthful conditions of the mucous membrane of the tongue If the tongue is dry taste is much lessened or abolished Since the sense of smell plays a considerable part in the production of the sensations ordinarily referred to the sense of taste a patient who has acute coryza or bad colds may complain of a loss of taste Large doses of the bromides blunt the sense of taste

The lesions affecting the sense of taste may be basal meningitis tumors and in juries Ageusia may be one of the manifold symptoms of hysteria which with facial paralysis is the commonest cause of a partial loss of the sense of taste In each case associated symptoms must determine the diagnosis

PARAGEUSIA Purely subjective perversions of the sense of taste are usually in dicative of hysteria they may constitute the aura of epilepsy in the insane they may be hallucinations Various bad tastes frequently are a source of complaint of neu rotic patients The administration of certain drugs may give rise to disturbed taste sensations

Petechiae Small extravasations are occasionally seen on the buccal mucous membrane and when present may be caused by one of the grave anemias hemo philia purpura hemorrhagica or scurvy or they may be the hemorrhagic infarcts of ulcerative or septic endocarditis

Exanthems Erythema and Vesicles *The rash of measles is often seen* on the pharyngeal and palatal surfaces prior to its appearance on the skin and Koplik de scribed as absolutely characteristic of measles small red spots with minute blue white centers on the inner surface of the cheeks which disappear as the rash develops The angry redness of scarlet fever involving the entire oral and faucial surfaces is very striking A slighter redness is present in simple erythematous stomatitis and the rare occurrence of a salivary calculus with accompanying redness should be re membered Vesicles when present may be the beginning of an aphthous stomatitis herpes the eruption of varicella or smallpox pemphigus or recent burns

Pigmented Spots Ulcers and Sloughs Dark spots usually of various shades of brown occur in dark skinned races or in mixtures of white and black races All of those diseases such as Addison s disease which cause a darkening of skin color may be manifested by dark spots in the mouth

Small grayish ulcers with reddened margins which have begun as vesicles situ ated on the inside of the lips and cheeks and on the edges of the tongue are the lesions of aphthous stomatitis the common canker sores If removal of exudate on the gums buccal surfaces tonsils or soft palate reveals ulcerations the exudate may

be due to Vincent s angina White curdlike patches beginning on the tongue and spreading to the inside of the lips and cheeks and leaving if detached a normal or slightly ulcerated surface form the disease known as thrush Aphthous lesions of the mouth recurrent ulceration of the genitals accompanied by iridocyclitis followed by hypopyon constitutes Beçhet s syndrome The etiology is unknown

Salivation and Dribbling The amount of saliva which is secreted in 24 hours is under normal circumstances from 1 000 to 1 500 ml Ptyalism or hypersecretion of saliva in which this amount is greatly exceeded may occur in early pregnancy dementia praecox encephalitis rabies and sometimes attends the menstrual period The pain due to dental causes alveolar abscess or trigeminal neuralgia and the process of dentition itself will induce an excessive formation of saliva Dribbling of saliva may occur as a result of hunger and at times because of inability to retain saliva as during sleep

Stomatitis Stomatitis often accompanies infectious diseases such as scarlatina measles influenza typhoid and rickettsial fevers and botulism The blood dyscrasias and certain forms of avitaminosis may cause pathologic changes in the mouth During the eruption of the temporary or milk teeth there may be stomatitis From the standpoint of general medicine catarrhal gingivitis catarrhal stomatitis and so called aphthous stomatitis should be looked on as a complication of many and various disease conditions that are manifested in the mouth

Epidemic Stomatitis (So Called Foot and Mouth Disease) Epidemic stomatitis is a disease of cattle and goats that is highly communicable to man It is characterized by slight rise in temperature and by eruption of vesicles on the mucous membranes of the mouth The causative agent is a filtrable virus which is present in the fluid contents of the vesicles and is excreted from the body through the kidneys the salivary glands and the mammary glands The virus is transferred directly through the mouth Occasionally the virus may produce the disease by entry through wounds The period of incubation lasts from 3 to 10 days

In the early stages of the infection there may be slight fever chills headache muscular pains and general malaise These symptoms are followed by the characteristic manifestations which are swollen painful lips and a thick coated tongue showing imprints of the teeth Within 2 or 3 days yellow vesicles appear over the soft tissues of the mouth rarely on the hard palate There is profuse salivation In severe infections 2 or 3 weeks after the onset of the disease large vesicles may develop on the hands and sometimes between the toes The vesicles may increase in size and coalesce to form blebs Occasionally the vesicles are observed on the lower part of the nose and may extend to involve the eyelids

Acute Herpetic Gingivostomatitis Acute infectious gingivostomatitis aphthous stomatitis catarrhal stomatitis ulcerative stomatitis Vincent s stomatitis are the cognomens of this disease

This is a common disease of young children due to the virus of herpes simplex There are fever irritability red swollen gums a vesicular eruption on the mucous membranes of the mouth oral fetor and local lymphadenopathy A constitutional reaction is always present but varies in severity a high fever temperature from 104 to 105 F (40 to 40 5 C) is common

The commonest finding is a characteristic involvement of the gums which varies from a thin red line along the dental margin to extreme redness and swelling of the gums In the rare infant infected before the eruption of teeth the gums escape involvement Lesions in the rest of the mouth when seen very early consist of a few to many small vesicles which soon rupture leaving grayish lesions of varying size single or confluent that may become ulcers The tongue and cheeks are most commonly involved

In some patients the tonsillar region is affected early and the disease resembles acute tonsillitis of bacterial origin Submaxillary lymphadenopathy of varying degree

is extremely common The disease lasts from 10 to 16 days regardless of therapy Pain tends to disappear within about 1 week from onset or about 3 or 4 days before the lesions heal Scarring is absent or very slight Swelling of the gums and lymphadenopathy among the first signs to appear usually persist for many days after the ulcers heal

Recurrent Stomatitis Occasionally herpetic stomatitis recurs The local lesions are similar to those described in the foregoing paragraphs but there is a virtual absence of systemic reaction in patients so affected Virus can be isolated from the lesions and circulating antibodies are present at the onset of symptoms The distinction of such lesions from etiologically different but clinically similar types of recurrent stomatitis depends on isolation of the virus or on results of biopsy or both

Gangrenous Gingivitis or Noma Gangrenous gingivitis or noma is an acute progressive gangrenous process of unknown origin primarily occurring in children usually beginning about the buccal gingivae and spreading rapidly to the mucous membranes of the cheek Soon there is extensive sloughing of the adjacent soft tissues about the mouth The intense rapidity of the progress of the disease the absence of a line of demarcation within the inflamed area and the comparative freedom from pain are characteristics of noma The disease is attended with great exhaustion and almost invariably is fatal

Oral Diseases Due to Physical and Chemical Agents The oral cavity is frequently exposed to injurious agents The ingestion and inhalation of foreign substances into the oral cavity lead to an accumulation of chemical physical and bacterial irritants which are prone to settle along the gingival crevices about the teeth The gingival portion of the oral mucosa is in a constant state of low grade inflammation if not actual disease

The different types of structure ranging from the oral mucosa to the highly specialized and mineralized enamel account for the great variety of response that is displayed in the oral cavity from the influence of physical and chemical agents However there are other channels of entry of noxious agents into the body (see Diseases Due to Physical and Chemical Agents Chapter 19)

Schour and Sarnat record that localized abrasions of the enamel of the teeth are found in occupations in which workers hold various objects between the teeth while at work such as tacks nails pipes musical instruments and whistles Fractures of the enamel and traumatic effects on the periodontal structures also occur Dust of abrasives such as sand and cement or ordinary powders and pastes used to clean the teeth may cause abrasions A distinction is made between abrasion and attrition Attrition is the physiologic wear of teeth incident to age

Acid fumes deposited on the exposed portions of the teeth react with the enamel and decalcification results Similar action on the teeth results from protracted vomiting in which the hydrochloric acid from the stomach decalcifies the enamel The ingestion of acid carbonated drinks accomplishes more slowly the same end results of decalcification and often caries

A high incidence of tooth decay and caries has been reported among workers who are exposed to sugar dust The sugar dust is supposed to be deposited along the labial gingival surfaces of the crown where it stagnates and induces abnormal fermentation and with the aid of bacteria induces acid production and decay of the teeth

Green staining of enamel pigmentation has been observed among copper workers Among silver workers there may be a black staining and there are black deposits at the necks of the teeth Staining may also be seen on the teeth of patients who ingest liquid iron compounds without using a glass drinking tube The green stain frequently observed on the mouth and gums of children is due to chromogenic bacteria

The exogenous gingival pigmentation of occupational origin is distinguished from

the endogenous brown melanin of the gingivae that is seen normally in the Negro and the mulatto and in white persons who have Addison s disease This melanin is situated in the basal layer of the epithelium whereas coal dust and metal dust accumulate on the surface and penetrate to various depths of the epithelium and sometimes even the dermis The sulfides of heavy metals are found only in the dermis but become visible as lines about the gingivae Certain common heavy metals produce the pigmentation of the gingivae that usually but not always follows the ingestion or inhalation of the metal That the ingestion inhalation or injection of lead or other heavy metal is not sufficient to produce a line is well illustrated in lead poisoning which follows the injection of lead for the treatment of metastatic malignant disease The essential factor is the presence of chronic gingivitis Lead absorption does not cause inflammation but rather inflammation brings out the lead line The lead line is manifested as faint mottled patches of black at the edge of the gum a fine black line along the entire gum margin or a black line wider along the margin of the gums Patches of black or blue black pigment occur on the cheeks The gingival lines produced by the heavy metals lead mercury and bismuth cannot be differentiated one from the other

Once a poison such as lead and mercury enters the blood stream it is precipitated as the heavy sulfide in the colon and at gingival margins or into the oral tissues

The alveolar processes and jaws may be the first structures affected by blood borne agents This is exemplified in yellow phosphorus poisoning in which the phossy jaw is a characteristic finding in mercurial poisoning in which the teeth become loose and are eventually exfoliated and in radium poisoning which was first diagnosed because of toxic effects on the jaw bones

In the early stages only the alveolar bone is affected In mercury poisoning the disease process may be limited to the alveolar portion of the mandible and maxilla and rarely extends much beyond the level of the apexes of the teeth In the later stages however the osteomyelitis may include a greater portion of the jaws and is followed by sequestration

Lesions of the Mouth in Vitamin Deficiency Lesions of the lips gums tongue and buccal mucous membranes occur as manifestations of deficiency of ascorbic acid and of some of the fractions of the vitamin B complex These manifestations are seldom found in pure form

Oral lesions associated with deficiency of riboflavin begin as an oral pallor followed by maceration and superficial transverse fissuring in the angles of the mouth These lesions are a type or types of perleche A superficial denudation of the mucosa may occur and cause the lips to become abnormally red along the line of closure In addition to the cheilosis there are seen fine scaly desquamations on a mildly erythematous base in the nasolabial folds on the alae nasi in the vestibule of the nose and on the ears Associated with these lesions keratitis part of a riboflavin deficiency may be present

Stomatitis and glossitis appear early in the course of nicotinic acid deficiency disease and they are manifested by swelling and redness of the margins and tip of the tongue As the disease progresses the redness becomes more intense until the parts assume a fiery scarlet hue Ulcerations appear along the sides the tip and the under surface of the tongue and on the buccal mucosa opposite the molar teeth The ulcers may be covered with a thick gray exudate

The function of ascorbic acid is concerned with the formation and regulation of the intercellular matrix When deficiency exists gingival lesions or scurvy occurs The essential lesion of scurvy is a vascular disturbance which results in swollen and boggy gums that bleed easily and may progress to actual ulceration Chewing is difficult and involvement of the alveolar bones causes the teeth to loosen and fall out The teeth themselves may be affected by the scorbutic process The dentin and enamel become coarse and defective with resulting rapid and early decay of the teeth

Hemorrhage into the buccal mucosa may result from damage to the vascular epithelium Lesions of the gums will not occur if the teeth have been extracted

Mucous Cysts Mucous cysts sometimes arise from the mucous glands of the mouth and of the tongue itself The anterior lingual gland often is affected As a rule these mucous cysts are small and are felt as hard rounded bodies beneath the mucous membrane Dermoid cysts occur in connection with the tongue but very rarely

Leukoplakia Leukoplakia occurs principally in men and is said to occur more frequently in men who smoke than in nonsmokers This statement however is questionable The condition begins with hyperemia following which a whitish granular surface develops This white granular surface may remain stationary for years or it may extend or malignant disease may supervene The beginning of a malignant process is suspected when the whitish areas begin to pile up and form wartlike structures Without any warning however by changes in the white areas metastatic nodules may appear in the neck and after their appearance it is found that the leukoplakia has developed into a malignant condition The lesions before malignant change develops are soft to the touch after malignant disease develops there is a considerable amount of induration and the border line which has previously been definite now becomes indistinct and hard

Leukoplakia is to be distinguished from the common benign condition of the buccal surfaces of the lips and anterior pillars of the tonsils characterized by multiple small yellowish spots These spots are enlarged sebaceous glands and the disease is known as *Fordyce s disease* Fordyce s disease is distinguishable from the Fox-Fordyce syndrome which involves the apocrine glands of the axillae and pubis The latter is under endocrine influence Lichen planus may resemble leukoplakia

SUMMARY OF ORAL CANCER

Carcinoma occurs within the oral cavity considerably less frequently than it does on the lip In 6 out of 10 who have intra oral cancers the tongue is involved 2 out of 10 will have cancer of the floor of the mouth 1 out of 10 has the lesion on the cheek and the last 1 will have it on the gums and the soft palate

Intra oral carcinoma tends to be more infiltrative in regard to both muscle and bone than does cancer of the lip and carries a correspondingly higher mortality rate It tends to produce earlier metastasis in the deeper cervical lymph node groups These metastatic lesions may be either contralateral or bilateral but rarely ipsilateral

Leukoplakia on the lip or in the mouth must be regarded as an important precursor of cancer A high percentage of tongue cancer is found to be associated with syphilis and leukoplakia Any ulcer fissure nodule or papillary growth existing for more than 2 or 3 weeks *should be investigated* Thickened patches of leukoplakia are suspected of being malignant when there is any change in their size or texture Of the various inflammatory lesions which appear like ulcerating malignant lesions are syphilis tuberculosis streptothrix and Vincent s angina A positive Wassermann reaction or a demonstration of fusiform bacillus is usually conclusive enough Biopsy studies are made of all of these lesions for the purpose of diagnosis

THE JAWS TEETH AND GUMS

THE JAWS

Familial Fibrous Swelling Various diseases of the jaws have been called fibrous dysplasia fibrous osteoma osteofibroma and ossifying fibroma which are now regarded as variants of a fibrous dysplasia Several members of a family may be affected with fibrous dysplasia of the jaws

The disorder is characterized by massive swelling of the cheeks and jaws beginning during the first three years of life Dentition is defective There are no associated symptoms or systemic manifestations unless the lower jaw becomes so large that it prevents closure of the mouth and interferes with respiration and speech

the endogenous brown melanin of the gingivae that is seen normally in the Negro and the mulatto and in white persons who have Addison s disease This melanin is situated in the basal layer of the epithelium whereas coal dust and metal dust accumulate on the surface and penetrate to various depths of the epithelium and sometimes even the dermis The sulfides of heavy metals are found only in the dermis but become visible as lines about the gingivae Certain common heavy metals produce the pigmentation of the gingivae that usually but not always follows the ingestion or inhalation of the metal That the ingestion inhalation or injection of lead or other heavy metal is not sufficient to produce a line is well illustrated in lead poisoning which follows the injection of lead for the treatment of metastatic malignant disease The essential factor is the presence of chronic gingivitis Lead absorption does not cause inflammation but rather inflammation brings out the lead line The lead line is manifested as faint mottled patches of black at the edge of the gum a fine black line along the entire gum margin or a black line wider along the margin of the gums Patches of black or blue black pigment occur on the cheeks The gingival lines produced by the heavy metals lead mercury and bismuth cannot be differentiated one from the other

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nence to the angles of the mandibles The reflex is obtained by tapping the lower jaw with a reflex hammer as illustrated in Figure 6 71

The temporomandibular syndrome is a temporal or bitemporal headache originating from the muscles of mastication and from the fibers of the temporomandibular joints (Williams)

Closure of the bite resulting in a dislocation of the stresses on the muscles of mastication may produce pain that is referred to the temporal region Tearing of the anterior portion of the joint capsule resulting from extraction of molars with the patient under general anesthesia may result in pain in the region of the joint Closure of the bite will produce a daytime headache while injury to the joint capsule is likely to produce pain at night when the mandible drops during sleep Closed bite rarely produces pain Capsular injury is a relatively commoner but still an unusual cause of pain in the head

Spasm Trismus or lockjaw a tonic spasm of the masseter and temporal muscles whereby the jaws are held firmly together may occur as a symptom of trismus neonatorum tetanus strychnine poisoning hysteria epilepsy or disease of the brain and is sometimes a reflex from dentition

Paralysis In paralysis the masseter and temporal muscles do not contract and there is inability to masticate on the affected side Paralysis of these muscles may indicate hemorrhage into the pons basal lesions (meningitis tumor caries) or neuritis

Pain and swelling from disease of the maxillae mumps quinsy or trichinosis may prevent the opening of the mouth and interfere with mastication

Ankylosis Ankylosis of the mandible is seen following traumatism or infection to the temporomandibular articulation A pseudoankylosis may be due to extra articular disease The ankylosis may be fibrous or osseous Unilateral ankylosis if it occurs before the fifteenth year presents a disfiguring shortening deformity of the affected side

Malignant Tumors The ulcerative type of malignant lesion may not be impressive on inspection On examination the border of the ulcer is hard and irregular in outline with varying degrees of fixation Usually the tumor involves one or more of the teeth very often the affected teeth have been pulled because the tumor has been suspected of resulting from diseased teeth The fungiform type of cancer may protrude as a solid mass the surface being smooth without noticeable ulceration

In *carcinoma* of the upper jaw there may be a degree of circumscription because an adamantinoma sometimes becomes malignant and protrudes through the gums Adamantinomas usually can be differentiated roentgenographically

Sarcoma occurs in the periosteum of the jaws and in the antrums The early sign of a periosteal sarcoma is the tumorous elevation that is diffuse The sarcoma most commonly is situated on the lower jaw In this situation it grows more slowly than in the maxilla Sarcoma of the antrum may develop more quickly than a periosteal sarcoma and may make itself manifest by expanding the bone then perforating it and spreading to the surrounding tissues Occasionally a sarcoma grows into the nostril and fills it this development being followed by a foul smelling bloody discharge Tumors of this type are highly malignant dangerous growths of poor prognosis

THE TEETH

Dentition The maxillae and the mandible carry the teeth The temporary or deciduous teeth appear with greater regularity as to order than as to time The permanent teeth arrive as follows first molars 6 years incisors 7 to 8 years bicuspids 9 to 10 years canines 12 to 14 years second molars 12 to 15 years third molars 17 to 25 years

Early Dentition Eruption of the teeth in advance of the usual dates is not of special significance

Roentgenograms show extensive multilocular rarefaction and pronounced expansion of the mandible The maxilla is also greatly expanded but the rarefaction is diffuse rather than multilocular No other bones are affected Near puberty the facial appearance improves with a progressive decrease in the size of the jaws by middle age the roentgenologic and facial abnormalities are minimal or absent Tissue specimens from the jaws show fibrous tissue containing varying numbers of giant cells

Osteomyelitis of the Jaws Owing to the variation in the structure of the upper and the lower jaws osteomyelitis is differently manifested in the two jaws

In the *upper jaw* the osteomyelitis may be localized about one or more teeth or may affect much of the bone However the process rarely extends beyond the alveolar process The upper jaw is affected by an extension from a tooth or from the frontal sinuses Rarity of extensive osteomyelitis in the upper jaw is due to a lack of marrow cavity in this bone

In children but rarely in adults infections in an antrum may be severe and the necrosis extend to the alveolar process and the hard palate

In local infections of the upper jaw well formed spicules of bone may be cast off at once or at intervals of varying length Only rarely is there an extension to a number of teeth so that a considerable portion of the bone is extruded In such instances the separated part is firm and is little eroded except in processes due to chemical intoxication particularly mercury

The *lower jaw* may be affected by an acute process simulating the acute osteomyelitis of the long bones The localized necrosis due to local infections may remain as such

Local infections of the lower jaw follow infection about the teeth and frequently are associated with the extraction of teeth The wisdom teeth are the chief offenders The masseter muscles become infiltrated or reflexly spastic so that the patient cannot open the mouth The edema may involve the submaxillary region and in severe cases suppuration in the deeper planes may occur even to the spread of the infection down the carotid sheath to the mediastinum Spreading to adjacent teeth may occur The removal of the bacterial focus may allow the infection to subside Infections following injury by certain drugs notably mercury arsenic and antimony may lead to more extensive destruction Even the local use of arsenic in the preparation for filling of teeth may lead to extensive destruction even to the loss of a part or all of the alveolar process The involvement of the soft parts in local bone infections is usually limited to a local abscess which opens about the gum or beneath the jaw

The marrow cavity in the lower jaw is limited Its blood supply is enclosed in the inferior dental canal In cases in which the inferior dental artery is occluded there is *destruction of a large part or all of the lower jaw* but usually the alveolar process alone is involved resulting only in the loss of a large part or all of the teeth Accompanying the involvement of bone there may be chills fever pain and swelling of the jaw The infection may extend to the neck In such cases wide suppuration of the soft parts of the neck is usual and extension down the carotid sheath into the mediastinum may follow In the severer cases death from generalized sepsis may occur

Owing to the density of the bone of the lower jaw the necrotic bone is extruded more slowly than in the long bones so that many months even years may be required to complete the process of separation

Defective Mobility *Inability to open or close the mouth may be due to spasm* or paralysis of the muscles of mastication The masseter temporal and other muscles concerned in the act of chewing are innervated by motor fibers from the fifth nerve (trigeminus) and spasm and paralysis of these muscles indicate some interference organic or functional with the action of this nerve

The jaw muscles may be tested by desiring the patient to clench the teeth If the muscles are intact lateral ridges are thrown up across the face from the malar emi

Early extensive or rapid dental caries may be due to rachitis but occurs also in pregnancy diabetes and chronic phosphorus poisoning Usually however it is of unknown cause The influence of carious teeth as a source of irritation or infection causing bad breath and adenitis is obvious

Grinding of the Teeth Gritting or grinding of the teeth during sleep does not necessarily indicate worms It occurs in children and in neurotic patients and other adult persons who are affected by pain and who for any reason sleep uneasily

Infections Infections of the teeth often originate from deeply extending dental caries Such infections are usually confined to the roots of the teeth and are defined as apical abscesses They are rarely larger than a few millimeters in size

The infections may not give symptoms In some instances they occur after the tooth has been rendered nonvital by the dentist but more frequently they occur spontaneously Pain in the affected tooth and swelling and tenderness localized over the area affected are the common manifestations Infections of a more acute character about tooth roots may involve a greater or lesser extent of the adjacent bone and thus cause an osteomyelitis

Tumors of Dentigerous Origin *Dentigerous cysts* begin insidiously The patient s attention is not attracted to the presence of the cyst until the expanding jaw produces a deformity These tumors can be demonstrated on roentgenologic examination They may become very large and may involve a greater part of the bone making surgical removal almost impossible because of the softness and fragility of the bone

Odontomas Odontomas tumors of toothlike structure are closely allied to dental root or radicular cysts Although they represent a more mature tissue development they often complicate radicular cysts A dense thickening of the jaws represents the physical findings which point to the occurrence of odontoma The diagnosis is confirmed by roentgenologic examination

Adamantinomas These are epithelial tumors of the jaw originating from epithelial remnants of the enamel They occur in young persons and produce expanding bone tumors which are first demonstrated by roentgenologic examination Under ordinary circumstances adamantinomas are noninvasive and nonmalignant tumors

THE GUMS

A *narrow band of bright red inflamed gum tissue* surrounding the neck of the tooth primarily on the buccal surface is a characteristic observation in all species of mammals It is not a recent disease nor is it the result of so called present day perverted methods of preparing and seasoning foodstuffs Mankind now enjoys the greatest variety of the most wholesome foods that has ever been available to any animal This narrowed band of inflamed gum tissue more pronounced on the buccal surface bleeds readily on slightest injury and is comparatively free from pain *Gingivitis* is a more profound infection of the gums than this reddened tissue surrounding the tooth It is an extension of this normally reddened margin

Pallor of the gums is noticeable in all forms of anemia *Red and spongy gums* may be caused by carious or illkept teeth attended with abundant tartar Spongy red and bleeding gums perhaps ulcerated occurring in artificially fed infants may be due to scurvy A similar condition in an older person may be the result of scurvy or combined vitamin deficiencies

Swollen or spongy gums are met with in many debilitating disorders such as leukemia pulmonary tuberculosis diabetes and purpura Ulceration along the line of the gums rarely extending to the cheeks or tongue is particularly characteristic of ulcerative stomatitis Finally localized or general redness and swelling may attend the eruption of the teeth and poisoning by chemicals and heavy metals

Diseases of the Gums *Acute Ulceromembranous Gingivitis* (*Trench Mouth*) This disease often is called Vincent s infection or trench mouth The dis

Delayed Dentition The most important cause of delay in dentition after the age of 15 years is rachitis together with other conditions or diseases involving mal nutrition particularly those occurring in the first 5 or 6 months of the infant s life The delay if observed is more significant of the past than of the present status A late appearance of the teeth may be indicative of cretinism

Difficult Dentition There has been much difference of opinion with regard to the influence of teething in causing disease

Anomalies of Dentition There may be a congenital absence (anodentia) of some or all of the teeth or a production of more than the normal number Supernumerary teeth in abnormal locations (for example the palate) arise from ectopic tooth primordia which have been displaced A third dentition has rarely occurred Sometimes fourth molars develop behind the wisdom teeth Teeth have been observed in which owing to a defect of the enamel organ the enamel was wanting Imperfect teeth are frequently associated with harelip Epithelial remnants of the dental lamina may give rise to cysts of various kinds situated in the gum

Notched Teeth If the permanent upper central incisors are somewhat rounded and peglike tapering from gum to edge with a single shallow and biconcaved notch in the edge it may be evidence of inherited syphilis (Hutchinson s teeth) These teeth are likely to be small to be placed somewhat irregularly, and to stand apart from one another If keratitis and disease of the middle ear coexist a presumptive diagnosis of syphilis can be made

Dentated Furrowed or Pitted Teeth If the edges of the teeth are *dentated* malnutrition is likely to be the cause *Grooves* or *furrows* running transversely across the teeth are indicative of an acute illness during infancy or childhood sufficiently severe to interfere with nutrition of the teeth *Pitted* teeth the molars exhibiting the greatest changes are caused by exposure to fluorine in the water

Loosened Teeth Loosening of the teeth in their sockets is associated with spongy ulcerated or bleeding gums and is often a familial manifestation Movability of the teeth usually is due to prolonged fevers pyorrhea alveolaris old age mercurial stomatitis scurvy purpura haemorrhagica phosphorus poisoning or gangrenous stomatitis

Caries The cause of dental caries is controversial Some believe that the structure of the tooth and metabolic and nutritional factors are the primary causa tive factors in tooth decay Others believe that local factors in the mouth such as bacterial acids the physical state of the food mechanical abrasion and a lack of protective action of saliva are the causative factors of tooth decay

A clear relationship between salivary amylase and the incidence of dental caries seems to exist In persons without dental caries hydrolysis of starch by saliva re quires approximately 45 minutes for completion The saliva of those with 4 to 6 cavities will complete the hydrolysis more quickly Without exception the rate of starch hydrolysis increases in direct parallelism with the number of *cavities*

The presence of fluorine in drinking water seems to decrease the number of cavi ties (see Fluorine Poisoning Chapter 19)

Recent investigations cited by Turner and Crane appear to support the impor tance of local factors particularly bacterial acids as a primary causative agent in tooth erosion Furthermore current studies emphasize the relation of *dietary carbo* hydrate intake to the formation of acids by the oral bacterial flora The restriction of the carbohydrate intake has been found repeatedly to reduce the oral *Lactobacil lus acidophilus* counts and to arrest the development of carious lesions Since *Lacto bacillus acidophilus* is a vigorous acid producing organism the inference is that the production of organic acids particularly lactic acid by this microorganism is re sponsible for erosion of the teeth Excessive amounts of acids in the mouth can cause dental erosion, this has been shown in experimental animals ingesting certain car-bonated beverages or dilute acid solutions and in patients taking excessive amounts of lemon juice for therapeutic purposes

will not cure the lesion If the lesion continues after the tooth has been pulled it should be suspected of being malignant

THE TONGUE AND THE FLOOR OF THE MOUTH

THE TONGUE

Inspection of the dorsal surface of the tongue is a part of the clinical examination which seldom was neglected by earlier physicians Perhaps never has so much been said about so little as in the nonsense written about appearances of the tongue as presenting evidence of diagnostic significance in many different diseases In general the tongue should be investigated top and bottom with reference to its color dryness and size palpated for lesions and tested for its mobility Occasionally a worthy bit of diagnostic information will thus be perceived concerning the physical status of the tongue itself

The mucous membrane which covers the tongue is modified skin Over the dorsal surface of the tongue are papillae The filiform papillae are the smallest and cover the anterior two thirds of the tongue The fungiform papillae are larger and are scattered on the dorsum sides and tip of the tongue and are set among the filiform papillae The circumvallate papillae seven to twelve in number form an anteriorly curved row at the base of the tongue In the median line posterior to the curved row of circumvallate papillae is the foramen cecum which is the upper extremity of the remains of the thyroglossal duct It is sometimes patulous for a short distance

On the posterior portion of the tongue behind the circumvallate papillae on each side of the median line is a mass of adenoid tissue which forms the lingual tonsil

In the middle of the dorsum of the tongue is a furrow caused by the septum binding the middle of the tongue down and allowing the muscles to rise on each side

Anomalies Reduction or absence of the tongue referable to developmental arrest and bifid and trifid tips through persistence of the unfused apical components are recorded

The commonest of all deformities of the tongue observed soon after birth is tongue tie a fixation of the tip of the tongue to the floor of the mouth by a frenum which is too short When the fixation is extreme the infant may not be able to nurse until the frenum is cut enough to loosen the tip of the tongue

Color and Pigmentation The color of the tongue as distinguished from the color of its coating is of some diagnostic value The most frequent abnormal coloration of the tongue results from either food or drink The tongue is pallid in anemia and bluish in cyanosis A bright red tongue may be due to the exanthems particularly in the early stage of scarlet fever or to inflammation of the tongue itself (glossitis) it differs from the darker reddened tongue of the deficiency states Petechiae ecchymoses and infarcts may be found on the tongue where they have the same significance as if found elsewhere Dark purple or blackish deposits of pigment may indicate an admixture of racial characteristics or an old glossitis or may constitute the discolorations of Addison s disease In jaundice the tongue is yellowish with the yellow tinge particularly marked on the under surface of the tip A series of clearly outlined yellowish white spots along the edges of the tongue is characteristic of xanthelasma

Uniform discolorations of the tongue and its coating may be caused by the ingestion of various substances corrosive or noncorrosive Ammonia bichloride of mercury and sulfuric carbolic and oxalic acids whiten the tongue hydrochloric nitric and chromic acids produce a yellow color Potassium and sodium hydroxide redden the tongue Among the noncorrosive drugs and foods bismuth charcoal and iron stain the tongue black wine fruits and berries color it red or purple and rhubarb tincture of opium tobacco licorice and chocolate stain it brown or black

Cross estimated that discoloration of the tongue constitutes at least 30 per cent of all oral reactions in patients taking penicillin orally The discoloration appears as

ease is caused by invasion of a mixed infection primarily represented by *Fuso bacterium plauti-vincentii* in association with *Borrelia vincentii* At the onset the patient is aware of an uncomfortable feeling and salivation and the tips of one or more of the interdental papillae on the buccal surface begin to swell They assume a livid grayish color and become the sites of small flat punched out ulcers that undergo necrosis The necrosis may destroy the gingivae down to the alveolar process A strip of brown colored membrane may cover the affected gingival margin Removal of the membrane leaves a bleeding surface The inflammation spreads rapidly but only in severe cases is the palatal surface of the gingivae affected Contact ulcers form on mucous membranes of the cheek or on the borders of the tongue The disease may spread posteriorly to the tonsils The mandibular lymph nodes are painful and enlarged There may be fever the temperature up to 104 F (40 C) The sense of taste may temporarily be completely lost

Trench mouth must be differentiated from avitaminosis scurvy pellagra and blood dyscrasias such as leukemia Although there may be extensive and widespread destruction of the gum margins the prognosis in trench mouth is good

Pyorrhea Alveolaris There is no known systemic disease that will cause pyorrhea alveolaris but there are two factors that produce the condition an ab normal occlusal stress on the articulating tissue of the infected teeth and coexisting disease that lowers the vitality of this articulating tissue Pyorrhea alveolaris always presents deep pockets containing suppurating granulation tissue with sooner or later loosening of the teeth The patient usually presents himself for general examination to find the cause of receding gums In the aged the periodontium often is smooth and atrophied This atrophy however may occur at an early age the condition should not be mistaken for pyorrhea alveolaris

Abnormalities and Tumors Hypertrophy This is a slowly developing proc ess and usually a great amount of deformity is present before the patient is aware that the gums actually are getting larger There are by the time awareness is aroused a nodulated enlargement of the gums and an irregular arrangement of the teeth On palpation the nodulations are densely hard The mucous membrane is not affected

Diffuse Fibromatosis of the Gingivae Diffuse fibromatosis of the gingivae may be hereditary No symptoms may occur until the fibromas become malignant and cover both sides of the alveolar processes of both jaws and even cover the crowns of the teeth By this time the teeth themselves may be crowded out of posi tion The surface of the fibroma is light pink and is often roughened by papules and ridges of proliferative gum tissue The tumors often are large enough to distort the facial contour

Epulis An epulis is a fibrous tumor of the gum An epulis on the lower jaw is small and of slow growth An epulis of the upper jaw adjacent to a molar tooth or a bicuspid tooth is usually large when first seen The surface of the tumor has the color of the adjacent mucous membrane The mucosa is usually continuous over the surface of the tumor Occasionally epulides are bluish red and vascular in ap pearance Epulis recurring after one or two removals may become malignant

Granulomas of the Alveolar Borders Dental granulomas develop from sites of irritation and are the result of infection The usual history is that the granuloma or tumor has developed within recent weeks within one or two months at the most The tooth on which the granulomatous tissue forms may be free of disease but usually the deposits on its surface irritate the adjacent gum

These lesions begin as soft growths of the gum adjacent to a tooth or several teeth If one tooth is involved the granuloma usually is not a large one As these tumors grow older they become harder and are usually less red than the surround ing tissue Removal of the tooth unless the granulomatous tissue also is removed

plies the muscles which fix and depress the hyoid bone in chewing and swallowing

If the cortical connections of the nucleus are involved there will be paralysis but no atrophy or if atrophy is present it is slight Contraction of the tongue due to the absorption under treatment of a lingual gumma may appear somewhat like paralytic atrophy

Spasm Spasm of the tongue is a rare symptom It may be tonic in which case the tongue is contracted and rigid or clonic in which case the tongue jerks and twitches irregularly or is protruded and retracted rapidly The movements usually involve both halves of the tongue but may be unilateral

When clonic spasm of the tongue exists it usually is a part or symptom of chorea hysteria or epileptic convulsions or is associated with mimic tic Stuttering is a spasmodic affection of the lingual muscles A curious disorder reported in those who use the voice incessantly for example solo singers is a spasm of the tongue on attempting to speak a condition analogous to writer s cramp Lingual clonic spasm is an occasional symptom in disseminated sclerosis and general paralysis Reflex irritation of the fifth nerve has been held responsible for lingual spasm and in some instances disease of the central nervous system produces it

Tonic spasm of the tongue is indicative of hysteria or reflex irritation through the fifth nerve occurring in nervously weak and debilitated persons It may coexist with tonic spasm of other voluntary muscles

Tremor A coarse tremor or trembling of the tongue is most frequently symptomatic of chronic alcoholism and is present in delirium tremens it usually is present in brominism and it may exist as a part of paralysis agitans A fibrillary tremor or fine twitching of the muscular bundles of the tongue may be seen in disseminated sclerosis in general paralysis and in bulbar paralysis when the lingual muscles begin to atrophy The tongue trembles in neurotic persons particularly when they are excited or agitated

Paralysis If one side of the root of the tongue is higher than the other and if when the tongue is protruded it deviates toward the same side unilateral lingual paralysis exists There are likely to be some interference with speech and a slight difficulty in chewing and swallowing If the tongue lies motionless in the floor of the mouth the patient being unable to protrude it and the functions of speech mastication and deglutition are greatly impaired there is total lingual paralysis In both cases there may or may not be atrophy and fibrillary twitching The absence of atrophy indicates a cortical or supranuclear lesion the presence of atrophy indicates a nuclear or infranuclear lesion

Lingual paralysis with atrophy of either one or both sides of the tongue according to the situation or extent of the lesion may exist as a symptom of the following conditions general paralysis chronic lead poisoning embolism or thrombosis of the vascular supply of the basal nuclei basal meningitis or tumor of the base syphilitic disease or other disease of the bone at the base of the skull or of the first cervical vertebra tumor of the upper portion of the spinal cord and wounds of the neck

Inflammations Inflammations of the tongue are likely to be diffuse Their cause often is obscure Lingual inflammations are characterized by pain burning and swelling These symptoms may remain for days and gradually subside without leaving a trace On examination the tongue is red and thick the thickening being evident on palpation The increased heat of the tongue is readily discernible by the palpating fingers If abscess formation has occurred a fluctuant region may be located on palpation

The tongue may appear enlarged when the floor of the mouth becomes inflamed Examination reveals edema under the tongue or under the chin Suppuration with abscess formation may follow

early as 2 days as late as 9 days and on an average of 4 days following the use of penicillin

The Black Hairy Tongue Black hairy tongue is an uncommon disorder and its cause is unknown It is thought in some instances to be due to a congenital abnormality which develops in later life It is a pigmentation and involves principally the back of the tongue the central furrow and portions of the pads leaving the sides and tip unaffected The color varies from yellowish brown to black

Histologically the filiform papillae are hypertrophic and hyperkeratotic and are the origin of hairlike filament that may grow as long as ½ inch (1 27 cm) The papillae become densely matted and deposition of pigment takes place The pigment is considered to be derived from food wines tobacco iron and mercury and mouth washes that contain oxidizing or reducing chemicals (Wolfson)

Black tongue has also been caused by the filamentous mycelia of actinomycosis which have been cultured from the tongue

Dryness The term appearance used with reference to the tongue usually relates to the character of the coating which may be present or to the disappearance of the coating and particularly, to its degree of moisture The coating consists of accumulated epithelium microorganisms and food detritus Dryness of the tongue when not caused by mouth breathing or coma is of very considerable importance as an indication of general dehydration and prostration

A thin white even furring of the tongue is normal in many healthy persons particularly those who are in the habit of smoking or of drinking milk and in mouth breathers Such a coating is always present in moderately febrile states

A tongue covered with a white fur through which project greatly swollen and bright red fungiform papillae is seen most frequently in acute infections either generalized or limited to the tongue This is not necessarily pathognomonic of scarlet fever

Size Great enlargement of the tongue is easily determined A slight increase in size can be assumed to be present if the edges of the tongue are indented by the teeth the indentations existing only if the tongue is swollen beyond its normal limits

Macroglossia This is a rare lesion of obscure origin Usually in such cases an abnormally enlarged tongue is observed in a child soon after birth A tongue of this type seems to grow slowly and the whole organ is involved in the processes of enlargement The tongue is firmer on palpation than the normal tongue It may become so greatly enlarged that it lacks the capacity to move and there may be local ulcerations where it has rubbed against or hung over the teeth

Enlargement of the tongue and a feeling of stiffness in it may accompany a general or regional amyloidosis Enlargement of the tongue occurs in acromegaly and in myxedema If associated with inflammatory symptoms the enlargement is due to acute glossitis If associated with urticaria or angioneurotic edema it is allergic in origin Tumors of the tongue also are responsible for an irregular and sometimes great increase in its size

Shrinking or Atrophy The tongue is small after a profuse hemorrhage and may become noticeably lessened in size during starvation

In atrophy of the tongue a condition which is due to some affection of the hypoglossal or motor nerve of the tongue involving its nucleus or its peripheral portion the mucous membrane of the tongue is thrown into folds and the organ has a shrunken appearance The atrophy is always conjoined with paralysis and the paralysis and atrophy affect one or both lateral halves of the tongue accordingly as the lesion is unilateral or bilateral Unilateral atrophy of the tongue sometimes is associated with facial hemiatrophy Unilateral hypertrophy of the tongue sometimes occurs in congenital hemihypertrophy

Spasm Tremor and Paralysis The motor supply of the muscles of the tongue is for the most part derived from the hypoglossal nerve The same nerve also sup

or both sides of the neck may become involved from a malignant growth in any portion of the tongue Lymph nodal involvement is not always palpable

The diagnosis is made on the basis of the histologic structure of biopsy tissue The prognosis is usually unfavorable

THE FLOOR OF THE MOUTH

On turning up the tip of the tongue a fold of membrane the frenum linguae is seen extending from the under surface of the tongue to the floor of the mouth beneath In newborn children this fold of mucous membrane appears sometimes to be too short hence the term tongue tied This anomaly may persist throughout life if not properly treated

Between the teeth and the tongue parallel to the alveolar process is the sublingual ridge formed by the sublingual salivary gland On the median side of the sublingual gland are the lingual nerve and the submaxillary duct (Wharton s duct) and on each side of the frenum on the anterior aspect of the sublingual ridge is a papilla into which the duct opens Opening into the submaxillary duct or by a separate duct into the same papilla is the duct of the sublingual gland (the duct of Bartholin)

The Submaxillary and the Sublingual Salivary Glands These glands present infections and tumors almost identical with those observed in the parotid glands The submaxillary glands like the parotids may be absent or displaced or there may be accessory glands

Infections and Inflammations These disorders of the sublingual and the submaxillary salivary glands are not unusual occurrences The glands may be swollen during an epidemic of parotitis but commonly they are affected in fevers and in serious illnesses much as the parotid glands are affected Occasionally there is a low grade inflammation of the sublingual and submaxillary glands which causes swelling of the ducts and obstruction to the flow of saliva during a meal Soon after eating the patient becomes aware of a swelling under the chin along the inside part of the lower jaw A swelling of this type may be painful but it usually subsides within a few hours it may not recur or it may become rather annoying and recur at frequent intervals The swelling is due to obstruction of the duct by a tenacious secretion and subsequent edema and pressure

Stones Formation of sialoliths in the submaxillary ducts and the sublingual ducts is much commoner than in the parotid ducts

Salivary calculi usually originate within the gland structure in the form of small concretions which are carried along with the salivary current to become lodged in the gland duct Here they become enlarged by deposits of bacteria and phosphates Any of the salivary glands or ducts may be the site of calculous formations but the most frequent location is Wharton s duct after which in order of frequency the submaxillary glands the parotid glands and Stensen s ducts

The symptoms depend on the size and situation of the stone If it lies within the gland structure the patient may experience only a mild intermittent feeling of irritation and discomfort in the affected region together with moderate swelling If however the stone is in the duct the symptoms may be severe characterized by a sudden sharp pain when the salivary flow is stimulated and rapid and painful distention of the gland In the course of 12 to 48 hours an inflammatory reaction may occur due to the retention of infected secretions General malaise may ensue

The floor of the mouth may be dry and frequently no saliva will be found flowing from the obstructed duct when the patient is asked to suck a piece of lemon Palpation of the duct may express pus or reveal the location of the stone Pus may be found in the floor of the mouth together with swelling of the tissues along the course of the duct with deviation of the protruded tongue when the submaxillary gland is involved A probe passed into a salivary duct may fail to locate the stone If the calculus is large enough to cause obstruction and particularly when it con

Eczema Eczema of the tongue gives rise to a condition variously known as *geographic tongue* annulus migrans or wandering rash Ring shaped patches red and denuded of epithelium are seen on the tongue and under observation are seen to spread at the edges while healing at the center coalescing and forming irregular areas with curved outlines Usually there is little or no discomfort from this condition

Scars, Fissures and Ulcers Scars Scars on the tongue may be the result of healed ulcers, of chewing the tongue during attacks of epilepsy or bulbar palsy of accidental biting of the tongue during restless sleep or in careless chewing of food or of a fall when the tongue is between the teeth

Fissures Fissures of the tongue aside from the normal median longitudinal one also are often of normal occurrence especially in elderly persons Deep fissures of the tongue are seen in Mongolian idiots In other subjects fissures are often significant of chronic glossitis Very deep inflamed fissures are often due to glossitis and in an occasional instance are of syphilitic origin Fissures or losses of substance at the edges of the tongue are usually due to deficiency states or to rough or broken teeth

Leukoplakia lingualis (ichthyosis linguae leukoma leukokeratosis buccal psoriasis) is manifested by slightly elevated thickened patches of irregular shape not ulcerated white and smooth on palpation which sometimes show a tendency to fissure They may furnish in 1 case in 10 the starting point of an epithelioma

Ulcers Acute and painful ulcers of the tongue are often due to aphthous stomatitis less often to the ulcerative form of stomatitis In chronic superficial glossitis shallow red glazed denuded areas occur on the surface of the tongue Multiple ulcers gray indolent stellate and associated with enlarged cervical lymph nodes may be tuberculous and are almost always secondary to tuberculous disease of the lungs Somewhat similar multiple ulcers may be syphilitic but in such cases the anterior cervical lymph nodes are rarely enlarged Multiple small ulcers preceded by vesicles occur in smallpox varicella measles and erysipelas as well as in pemphigus herpes and eczema of the tongue

A single lingual ulcer with a hard base associated with enlarged cervical lymph nodes may be either the initial lesion of syphilis or an epithelioma When syphilis exists the age of the patient is usually less than 40 years The lesion is on the tip of the tongue is not very painful and improves under treatment A mucous patch may become ulcerated or a gumma of the tongue may break down forming a deep sore A single ulcer if opposite a rough or jagged tooth may be simply the result of continued trauma but the presence of an irritating tooth does not exclude epithelioma or syphilis

An *ulcer* on the *frenum linguae* may occur during the course of severe coughs such as pertussis from the thrusting of the undersurface of the tongue against the lower incisors habitually or during paroxysms of coughing

Tumors Benign Lingual Tumors Any kind of benign tumor of the tongue is uncommon in adults Papillomas small in size are the commonest of the benign tumors Fibromas cysts lipomas and hemangiomas may be encountered

Malignant Lingual Tumors Carcinoma of the tongue is commoner in men than in women and commoner in middle aged men than in either the young or the old

Carcinoma of the tongue begins on the surface of the tongue either by a change in the epithelial covering or in fissures or ulcers at its edges Some carcinomas of the tongue begin in leukoplakial spots a few begin as benign papillomas and still a few more begin in ulcers caused by broken teeth or defective dental structures A greater number however just begin as sore places and continue as enlarging ulcers

The lymphatic vessels from the mucous membrane and deep tissues of the tongue have apparently only a single outlet so that the lymph nodes on either side

fluids through the nose on attempting to swallow and if there is failure to pronounce correctly certain sounds for example the patient says beng for beg Inspection of the soft palate during the utterance of the long ah sound will show under normal circumstances that both sides of the palate arch upward If the whole palate remains motionless during phonation of ah there is bilateral paralysis if but one side moves paralysis is unilateral

Paralysis of the soft palate is most commonly due to diphtheritic neuritis or poliomyelitis but is also caused by bulbar paralysis brain tumors basal meningitis and vertebral caries

Anesthesia of the Palate and the Fifth Cranial Nerve *Anesthesia* of the hard and soft palates indicates disease involving the second division of the *fifth nerve*

The motor portion of the trigeminus supplies the muscles of mastication namely the two pterygoids the temporal masseter mylohyoid and anterior belly of the digastric

The sensory portion of the trigeminus affords sensation to the face conjunctivae nose frontal and maxillary sinuses teeth palate tongue part of upper pharynx external auditory meatus and the scalp as far back as the vertex It may or may not supply the sense of taste on the anterior two thirds of the tongue

The motor functions of the trigeminus are tested by having the patient clench the teeth firmly while the temporal and masseter muscles are palpated in order to observe their contraction Also have the patient open his mouth If the muscles on one side fail to contract and the opened jaw deviates toward the paralyzed side there is unilateral paralysis due to disease affecting at least the motor fibers of the inferior maxillary division of the trigeminus Spasm of these muscles constitutes trismus or lockjaw

The sensory functions of the trigeminus are examined in the ordinary manner Whether one or all of its divisions are involved may be determined by comparing the areas of anesthesia found with its distribution If there is complete unilateral anesthesia the patient when drinking can feel the contact of the cup or glass on one side only affording a sensation as if half the vessel were missing The sense of taste is partly conducted through the medium of the trigeminus There are certain inflam matory (herpes zoster) vasomotor (flushing pallor) and secretory (lacrimation salivation) disturbances which may arise from disease of the trigeminus because of the fact that it supplies the lacrimal and salivary glands with secretory fibers and is also the channel by which vasomotor fibers run The trigeminal nerve is involved in neuralgias and migraine Trigeminal anesthesia is seen in hysteria and in various forms of organic disease which to a greater or lesser extent involve the cranial nerves or the cranial nerve centers Areas of cutaneous anesthesias when the cutaneous nerves of the head and face are involved are shown in Figure 4 1

Tumors Slowly developing tumors of the palate are exostoses and ecchon dromas They are situated in the midline grow very slowly and cause no trouble except that the patient is conscious of their presence The appearance of the tumors is characteristic They feel smooth and hard unless they are myxomatous or malig nant

THE FAUCIAL TONSILS

The tonsils are oval and when normal in size project but little if at all beyond the pillars of the fauces They contain several recesses or crypts formed by the folding inward of the mucous membrane The walls of the crypts contain lymphoid tissue and mucous glands Extending from these crypts are pockets known as follicles These follicles may become sealed from inflammation which leaves palpably hardened areas in the tonsils

Acute Tonsillitis Tonsillitis is most frequently met with between the ages of 10 and 30 years The disease sometimes occurs in children less than 10 years of age and in adults of more than 50 years of age Tonsillitis accompanies many acute

tains calcium salts as its chief constituent it is usually demonstrable in a roentgenogram

Cysts The term ranula generally is applied to any cyst in the floor of the mouth that is due to obstruction of a duct of either a mucous gland or a salivary gland The cysts arise just behind the lower incisor teeth and may be due to involvement of the incisor gland (glandula incisiva) but most commonly they are due to obstruction of the excretory ducts of the sublingual glands Among ranulas have been included retention cysts as well as congenital inclusion cysts Ranulas must be distinguished from dermoid cysts The dermoid cyst has a thicker feel more firm on palpation and it often pits on pressure

Tumors Submaxillary tumors begin as small swellings beneath and medial to the border of the jaw just in front of the angle of the jaw As a tumor of this type becomes larger it may occupy a considerable region of the neck It is impossible to tell whether the growth springs from the submaxillary gland or the sublingual gland until the affected gland is identified

One of the common tumors of the submaxillary gland as of the parotid gland is the mixed tumor The hard but elastic feel of these growths is characteristic The tumor is easily palpated by one finger of one hand on the floor of the mouth with the other hand under the chin If the tumor has become malignant it is fixed more closely in its position

Carcinoma of the Floor of the Mouth This lesion usually is an ulcer with hard borders It often is found accidentally before it gives the patient any symptoms Some growths of this type are small rounded masses circumscribed and somewhat movable large tumors may form however filling the whole floor of the mouth and bulging into the neck

THE PALATE

The superior and lateral boundaries of the oral cavity and the differentiation of the nasal and oral cavities are the result of fusion of the two lateral maxillary processes of the first branchial cleft and the medial frontonasal process which descends from the anterior part of the head in the embryo Failure of fusion of these processes in part or entirely in embryonic life results in one or another variety of cleft palate *Cleft palate is discussed with cleft lip*

The anterior and posterior arches of the palate and the uvula are plainly visible The anterior pillar extends from the soft palate to the tongue and is formed by the palatoglossus muscle The posterior pillar extends from the soft palate downward to the sides of the pharynx and is formed by the palatopharyngeus muscle In front of these arches and extending from the roof of the mouth opposite the posterior edge of the last molar tooth downward to the posterior edge of the alveolar process of the lower jaw is an elevation of the mucous membrane which shows the line of junction of the hard and soft palates

Torus palatinus is a protuberance of the hard palate at the line of closure of the maxillary sutures These swellings are unimportant clinically They are often of concern to the patient

The anterior three fourths of the roof of the mouth is formed by the hard palate and the posterior one fourth by the soft palate

Vesicles arranged in annular shape on the soft palate or perhaps on the pharyngeal wall and attended with a disproportionate amount of pain are viral herpes of the same sort that occurs elsewhere on the mucous membranes of the mouth When these vesicles rupture small ulcerations follow and constitute the commonest acute ulcerations of the palate Chronic ulcers of the palate are often of syphilitic origin

Perforation of the soft palate or its *adhesion* either partial or complete to the posterior pharyngeal wall may be the result of any severe infection or due to syphilitic ulceration or diphtheria

Bilateral paralysis of the soft palate is to be suspected if there is regurgitation of

portion of the membrane or of a swab from the surface of the tonsils The presence of Klebs Loffler bacillus (*Corynebacterium diphtheriae*) when culture is made on the proper media indicates diphtheria

In regard to the relationship of acute tonsillitis and scarlet fever see Strepto coccal Infections in Chapter 17

In cases of uncomplicated tonsillitis the outlook is always favorable The disease runs its course in about a week Peritonsillar abscesses or quinsy may occur and when they do develop they greatly increase the discomfort of the patient In peri tonsillar abscess or quinsy the prognosis is more guarded especially because of the possibility that the more virulent infection may produce septicemia or Ludwig s angina

Corynebacterium diphtheriae (Klebs Loffler Bacillus) and Diphtheria The causative organism of diphtheria is of the order Eubacteriales Buchanan family Coryne bacteriaceae Lehmann and Neumann genus *Corynebacterium* Lehmann and Neumann and species *Corynebacterium diphtheriae*

The optimism that prevailed during the first three decades of the twentieth century with regard to the successful therapeutic control of diphtheria yielded to pessimism and perplexity when numerous instances of malignant diphtheria occurred in central Europe for these cases were wholly refractory to routine methods of serum therapy Later the instances of *malignant diphtheria* proved to be a special type due to *Corynebacterium diphtheriae gravis* Since this division was made the name *Corynebacterium diphtheriae mitis* has been employed to designate the nonmalignant strain

The extracts of the gravis strain produce necrotizing and edematous lesions resembling the local reactions to potent staphylococcal or streptococcal toxins The main product from gravis strains is an endotoxin analogous to the Duran Reynals spreading factor formed by certain invasive strains of pyogenic cocci The severe lesions of malignant diphtheria are due to the synergic effects of classic diphtheria toxin with gravis endotoxin The gravis strains of diphtheria may account for the exceedingly large quantities of antitoxin required for the successful control of the symptoms in an occasional patient who has diphtheria

There are conferred active immunity and passive immunity to diphtheria Active immunity now is conferred by the use of especially prepared toxoid solution of which one injection produces immunity

The diphtheria organisms are transmitted by droplets food handling of articles in common use such as toys and by contact directly or indirectly through clothing The germs can live for weeks on toys and clothing and on various household utensils that are not in common use

The carrier rate increases with the beginning of cold weather The presence in the nose of a subject of bacilli that appear morphologically to be those of diphtheria is not enough to justify giving the person the stigma of carrier and enforcing isolation for treatment The bacilli should be isolated and their virulence proved bacteriologically before the subject is labeled a diphtheria carrier and before isolation is advised

Susceptibility of the individual to diphtheria can be determined by tests If suscepti bility exists prophylactic treatment can be given that usually suffices for control

Schick Reaction The blood of the majority of normal adults contains a small amount of diphtheria antitoxin a fact which probably accounts for resistance of many persons to diphtheria The presence of the antitoxin may be easily detected by means of the Schick reaction A standardized diphtheria toxin is diluted in physiologic salt solution so that 0 1 ml of the solution contains 1/50 of the minimal lethal dose for a guinea pig This dose is injected intracutaneously If the blood of the subject tested has less than 1/30 unit of antitoxin per milliliter a positive skin reaction appears in from 24 to 36 hours and persists for 4 or 5 days or more A positive reaction consists in a slight infi tra tion of the skin surrounded by a red areola 1 to 2 cm in diameter Positive reaction indicates susceptibility negative reaction indicates natural immunity to diphtheria The naturally immune person however may harbor the bacilli and be a carrier it follows therefore that many carriers may give negative reaction

Negative reactions to the Schick test are obtained in 9 out of 10 of the newborn but become less frequent in children up to the second to the fifth year As children grow

illnesses It is present in the initial stage of measles and scarlet fever It is frequently present in diphtheria and in secondary syphilis Water and milk may carry infective organisms for tonsillitis Tonsillitis is sometimes infectious and not infrequently epidemic Hemolytic streptococci often in pure culture have been found during epidemics of acute tonsillitis in military camps and in civilian practice The diplococcus of pneumonia is said to be responsible for less than 10 per cent of cases of acute tonsillitis and to the *Hemophilus influenzae* organism (bacillus of Friedlander *Klebsiella pneumoniae*) is attributed a very large percentage of benign membranous tonsillitis

If the tonsillitis is limited to the catarrhal variety the surface of the tonsils shows the changes common to inflammation of the mucous membrane In other cases the so called lacunar variety the morbid changes are confined mainly to the crypts In the so called parenchymatous tonsillitis there is congestion of the whole tonsil

SYMPTOMS An attack of tonsillitis is not necessarily ushered in with symptoms referable to the throat There may be such symptoms as fever sometimes chills and fever the temperature reaching 102 to 103 F (38 9 to 39 4 C) and in severe infections as high as 105 F (40 6 C) In most instances of tonsillitis the local symptoms make themselves felt at once The illness begins with a feeling of fullness in the face and the throat amounting soon to sharp pain which extends up to the ear and is generally augmented on attempt to swallow There may be so much pain on trying to swallow that the patient quits eating and drinking The voice becomes thick and muffled the senses of smell and taste are suspended and there may be deafness from eustachian catarrh Headaches backaches and pains in the arms and legs are of common occurrence

EXAMINATION The lymph nodes at the angle of the lower jaw are swollen and in most cases are very tender so that the head is held stiffly The throat is often difficult to examine in those who have acute tonsillitis for the pain is intense when they try to open the mouth and consequently a view of the back of the throat is often facilitated by the use of a head mirror The inflammatory process extends to the anterior faucial pillars the uvula soft palate and pharynx where the evidence of pharyngitis may also be apparent The congested surface of the tonsils is often spread over irregularly with debris which occludes the orifices of the tonsillar crypts This debris may accumulate and as time passes may form a white grayish or dirty yellow membrane This membrane in contrast to that accompanying diphtheria is readily separated from the tonsil and does not leave a bleeding surface when it is removed The swelling produces obstruction of the circulation in the region of the tonsil and leads to edema in the more relaxed tissues on the posterior wall of the soft palate which may be seen hanging down in a translucent bag on each side of the uvula The soft palate may become so impeded in its action that the voice becomes almost unintelligible and the attempt to swallow liquids may be followed by regurgitation through the nostrils

Tonsillar abscesses are rare but when they do appear they discharge their purulent contents into the pharynx without ill effects

DIAGNOSIS The diagnosis generally presents no difficulty Acute follicular tonsillitis must be differentiated from diphtheria Diphtheria generally is more gradual in its onset and it is more likely to begin on one side or at least it does not start on one side and then develop on the other as does tonsillitis There are less pain and fever in diphtheria The mouth is opened more easily and the throat with the characteristic adherent spreading false membrane may be viewed more easily in diphtheria than in follicular tonsillitis The false membrane in diphtheria generally invades the soft palate and uvula and often spreads to the larynx postnasal space and the nose The younger the patient the more likely is the condition to be diphtheria In doubtful cases the final diagnosis is to be made after culture of a small

whereas in others there is a chronic parenchymatous hyperplasia in which the tonsil is soft and friable in still others there is a slow fibroid degeneration an increase in the connective tissue stroma In tonsils with fibroid degeneration there may be cyst formation or the deposit of calcareous masses

Of paramount importance is the fact that the size of the tonsil is not a criterion of an infectious disease Some of the largest tonsils give rise to the least local inflammatory trouble whereas small tonsils which have adhesions about their borders and the remains of tonsils which have been imperfectly removed surgically may be the source of serious illness of the patient The tonsil is more to be suspected as a septic focus when creamy pus can be expelled regularly from the fossa or when a congested appearance of the tonsil suggests an infection in the follicles themselves which has cut off communication with the surface Such conditions may be associated with permanent injection of the buccal surface of the anterior faucial pillar and sometimes with enlargement of the lymph node just behind the angle of the jaw On neither clinical nor bacteriologic examination of the tonsils before operation can it be decided that a tonsil is a focus of general infection even if hemolytic streptococci are obtained in pure culture from the tonsils The decision in regard to focal infection can be made only after the tonsils have been removed and the patient has returned to good health The return of good health is presumptive evidence that the tonsils were a focus of infection

Kaiser did not observe immediate benefits after the removal of tonsils in children but over a 10 year period following the removal of tonsils there were a reduction in the incidence of sore throat and a decrease in the susceptibility to scarlet fever diphtheria cervical adenitis first attacks of rheumatic fever and otitis media

SYMPTOMS In some children and adults there are very definite symptoms from enlarged tonsils The tone of the voice frequently is thick and there is a ring or resonance about it which can be distinguished from that caused by nasal or laryngeal obstruction Articulation may be indistinct If the patient makes a complaint it may be of a sensation of fullness in the throat Often there is a recurring eustachian catarrh with each bad cold The cervical lymph nodes are chronically enlarged There may be a foul breath or a disagreeable taste in the mouth

Hypertrophied tonsils which should be surgically removed are accompanied by a history of frequent attacks of tonsillar inflammation or of peritonsillar abscess There is interference with respiration by day or by night and occasionally an alteration of the voice is noticeable

The prognosis after the removal of the tonsils in those who have had repeated attacks of eustachian catarrh or inflammatory disease of the middle ear is guarded The removal of the tonsils in the presence of these diseases should be advised strictly on its own merits for treatment of the local condition which may be demonstrated No doubt at times general systemic benefits will be obtained from the removal of the tonsils but these benefits cannot be predicted

Singers or those who wish to be singers often want to know if removal of the tonsils will interfere with singing In those who are already singers there is certainly a risk that the instinctive control of the muscles used in singing may be temporarily lost to such a degree as to impart a peculiar tone to the voice which previously was prized However in those who are already singers after tonsillectomy a renewed study and practice will often result in a stronger and better voice if not one of precisely the same tone

Patients who are only beginning the study of singing can safely be advised to have the tonsils removed if removal is indicated on other grounds

Generally it may be said that the speaking voice undoubtedly is improved in all cases after removal of the tonsils although it may take a little time for the patient to relearn the complete use of the muscles which have been liberated by removal of the tonsils

beyond the fifth year negative reactions increase and in adult persons the same ratio of negative to positive skin tests prevails as in newborn children that is 9 out of 10 are negative

SYMPTOMS The period of incubation of diphtheria varies from 2 to 10 days but is usually 2 or 3 days The onset is with fever temperature 102 to 104 F (38 9 to 40 C) chilliness headache and pain in the back and limbs

There is nothing distinctive in the initial symptoms which vary according to the site of the specific inflammation Generally there are sore throat and dysphagia A croupy cough hoarseness or aphonia and above all evidences of progressive laryngeal stenosis constitute a dangerous involvement of the larynx If there is an extension of the membrane into the trachea and the bronchial tubes the breathing may become stridulous and dyspnea and cyanosis are manifest

Often the symptoms of diphtheria are mild with slight fever and little if any prostration In diphtheria gravis there are extensive and intense local changes the prostration is extreme the temperature is subnormal the face is ashy the pulse rapid and feeble Subcutaneous ecchymoses appear

Extraneous infections and symptoms of diphtheria are common A cutaneous wound may become infected and a pseudomembrane may form at the point of lodgment of the germs Diphtheria of the conjunctiva may occur primarily or by extension from the nose, of the external ear, from the discharge of a diphtherial otitis media of the mouth and lips by extension of wounds ulcers and the genitals by direct infection from contaminated hands instruments or dressings Diphtheritic paralysis may be a serious complication (see Chapter 15)

EXAMINATION There are swollen and tender cervical lymph nodes There may be apparent stiffness of the cervical tissues

The tonsils are swollen and covered with grayish or yellowish white membrane which is usually adherent and leaves a bleeding surface when forcibly stripped off By the second or third day the membrane extends to the faucial pillars perhaps also to the uvula and the posterior pharyngeal wall Under favorable conditions the symptoms subside and the throat becomes clear of membrane by the tenth day

If the inflammation extends to the nose there is an offensive often bloody discharge from the nostrils or epistaxis In certain instances there may be thick membranes in the nose containing diphtheria bacilli but constitutional symptoms are absent and the disease is relatively harmless (fibrinous rhinitis) On extension of the infection to the tonsils the membrane may appear punctate and be confined to the tonsils simulating an ordinary follicular tonsillitis or there may be a soft nonmembranous exudate on the tonsils alone or the disease may be so extremely mild that membrane or exudate does not appear the throat presenting only the redness of an ordinary catarrhal inflammation

On extension of the disease to the larynx the supraclavicular episternal intercostal and epigastric spaces are deeply retracted with inspiration and bulge with expiration Shreds of membrane may be coughed up If the stenosis is not relieved the child passes into a semicomatose state and death ensues after a few hours to several days

DIAGNOSIS The diagnosis is established by positive cultures from the throat for the Klebs Loffler bacillus in the presence of a membrane and other variable symptoms and signs as enumerated

Chronic Tonsillitis Chronic Enlargement of the Tonsils The cause of lymphoid hypertrophy of the tonsils is not known Generally the tonsils begin to enlarge at the age of 3 or 4 years They begin to shrink at about the age of 12 years and tend to diminish or to become much smaller at about the age of puberty Sex does not seem to have any influence on occurrence of tonsillar hypertrophy In a hypertrophied or an enlarged tonsil there is an increase in the normal structures In some the crypts are more distinctive and contain a variable amount of mucus

unilateral enlargement or a deep spreading ulceration on an enlarged tonsil from which an offensive sanguineous discharge issues is in all probability cancerous

Most malignant tumors of the tonsils are anaplastic epidermoid carcinomas arising in the mucous membrane covering the tonsil or lining a crypt Lymphosarcoma of the tonsils arises less frequently The malignant lesions arising within the tonsil itself are usually more rapidly growing and much more radiosensitive than are carcinomas arising on neighboring mucous membranes and invading the tonsil secondarily The diagnosis is made by biopsy study of tissue secured from the lesion or of the removed tonsil

THE NOSE

The nose is long short flat small or large The contour of the nose viewed in profile is formed by an upper portion of bone the bridge of the nose a middle of cartilage and a lower or tip also of cartilage The bridge of the nose slopes downward and forward and where it joins the upper lateral cartilage the line changes and slopes more sharply downward until the tip is reached Here the lower lateral cartilages bulge forward forming a rounded and often a projecting tip

The skin over the tip and alae is thick and adherent to the cartilages It possesses a comparatively scanty blood supply hence its liability to suffer from exposure to cold and it is a favorable site for ulcerations (lupus) and superficial epithelioma (rodent ulcer) Sebaceous and sweat glands are abundant The skin is often pock marked from acne Stiff hairs guard the inside of the nostrils The follicles of these hairs are often the seat of small furuncles or boils which are extremely painful owing to the tension caused by the congestion and the swelling which is restricted by the tissues being so firmly bound to the cartilages beneath

Anomalies and Deformities A nose with stenosed nares or an incomplete septum represents a persistence of the normal fetal anatomy A failure of the region between the nasal sacs to consolidate into a septum permits apical bifurcation or duplication of the nose A rare anomaly of the nose is cyclopia a tubular proboscis attached above the single median eye

The nose may be long and its midportion bowed forward with the tip turned down toward the upper lip Such a deformity is of developmental origin often hereditary Contrasted with the nose of this shape is the short nose with the external surface curved inward toward the face with the nares large and pointing outwardly and very obvious The shape of this nose too is of developmental origin often hereditary A comparable deformity less the forward opening of the nares is produced by surgical injuries and traumatic fractures of the bony part of *the nasal septum This deformity may be but is rarely due to syphilis* The nasal septum may become ulcerated and perforated and deformed so severely from the inhalation of toxic gases that the external contour will become deformed

In cretinism and myxedema the nose becomes coarse and broad It is broadened or distorted and displaced by growing tumors in the nasal cavities or in the adjacent facial bones

Rhinophyma and Redness Rhinophyma is a chronic hypertrophy of the skin of the nose and edges of the alae nasi The cause is rosacea and the condition occurs in middle aged or aging men

Chronic redness of the nose is due to dilated capillaries and if not normal it usually results from sunburn windburn or rosacea It is rarely due to chronic alcoholism

Small dilated blood vessels appearing over the tip and alae of the nose occur in different disease conditions especially cirrhosis of the liver In this disease the small nevi vascular spiders formed by small dilated vessels which are known to be minute arteries are a common although not a diagnostic manifestation The

Peritonsillar Abscess (Quinsy or Cynanche) Inflammation and formation of pus in the tissues adjoining but outside the tonsil is known as quinsy The condition is a peritonsillar abscess

The illness begins with fever and a chill which is followed by an acute pain behind the angle of the jaw and extending up to the ear and down the neck on the same side as the abscess Swallowing is so painful that often the patient will allow the saliva to run from the open mouth rather than endure the pain that attends an attempt to swallow or to expectorate Accumulation of secretions in the pharynx adds a rattling character to the heavy thick tone of the patient s voice The neck is held stiffly and the head is forward and inclined to the affected side The tongue is protruded with the greatest of difficulty Thirst often is intense Taste is lost early in the disease and the sense of smell is diminished The patient s breath becomes offensive and the patient is pale and anxious and soon is very much exhausted The temperature in the early stages of the disease may reach as high as 105 F (40 6 C) The abscess often ruptures through the tonsil itself into the superior tonsillar fossa or through the anterior pillar and soft palate With rupture of the abscess after 5 to 10 days the general symptoms cease at once and the local discomfort abates rapidly Some days or weeks are required before the patient returns to normal health

On examination there is tenderness over the whole side of the neck The lymph nodes at the angle of the jaw and along the sternocleidomastoid muscle are swollen and tender Diffuse tenderness also may be present in the submaxillary parotid or occipital nodes The isthmus of the fossae is considerably narrowed by the bulging of the anterior pillar and neighboring portion of the soft palate The swelling may be such that the tonsil itself which may not actually be swollen at all may not be visualized The surface of the tonsil and the abscess often is coated with a deposit of mucopus resembling a false membrane which is readily detached A valuable sign in the diagnosis of a peritonsillar abscess is a loss of mobility of the soft palate on the affected side The tip of the uvula tends to point toward the tonsil around which there is an abscess Edema of the larynx is rare but if it occurs it may be of very serious import

A patient usually recovers from a peritonsillar abscess but the spontaneous bursting of a large abscess which floods the trachea with pus while the patient is asleep may cause death Hemorrhage from a peritonsillar abscess may occur as a primary or as a secondary event with or without surgical drainage of the abscess

Among the possible complications of quinsy are septicemia bronchopneumonia thrombophlebitis pyemia cellulitis of the neck seropurulent pleurisy meningitis suffocation from pressure secondary edema of the glottis and acute edema of the lungs

Tuberculosis In the adult tonsillar tuberculosis is of slight importance because on the whole it is only a secondary localization in an advanced pulmonary tuberculosis In children it presents as either a secondary localization to pulmonary tuberculosis or a form of pharyngeal infection Tonsillar tuberculosis may be latent and thus is detected only by histologic examination Tuberculosis of the cervical lymph nodes and tonsillar tuberculosis are frequently associated Every patient whose tonsils disclose tuberculosis should be kept under observation until it is clear that pulmonary tuberculosis is not present Tonsillectomy involves no great risk in the presence of tonsillar tuberculosis if the pharynx too is not deeply involved

Tumors There is a general belief that simple tumors of the tonsils are practically nonexistent This statement is generally true however papillomas adenomas fibromas angiomas dermoid tumors and occasionally lipomas are all seen in this region In addition to these tumors occasional keratomas mixed tumors and retention cysts and hardened areas of scar tissue are seen

Malignant tumors of the tonsils occur after 50 years of age In an elderly person

Discharges From the Nose Nasal discharges may be watery mucous muco purulent or purulent or bloody or composed of blood alone according to the nature and seat of the causal condition

A *watery discharge* marks the beginning of acute coryza vasomotor rhinitis hay fever the catarrhal form of epidemic influenza pertussis measles and typhus and other fevers In a few days a watery discharge often grows thick and muco-purulent Watery fluid may flow from one nostril during an attack of trigeminal neuralgia An occasional watery discharge with persistent obstruction of one or both nostrils may be due to nasal polyps A recurring flow of pus from one nostril particularly if brought on by lying on or leaning over toward the side opposite to that which is discharging is probably due to an antral abscess and the probability is increased if bad teeth are present In children the possibility of a foreign body should not be overlooked in all instances of nasal discharges not easily accounted for Finally irritating gases or powders will produce a smart watery flow

Bloody discharge may be significant of an impacted foreign body Coming on in the course of pharyngeal diphtheria and irritating the parts with which it comes in contact such a discharge may indicate nasal infection before membrane can be discovered in the nasal cavities An offensive discharge from both nostrils which may be accompanied with greenish gray crusts is symptomatic of atrophic rhinitis or may be due to syphilis caries or necrosis of the bones It may be a sequel of scarlatina The patient is usually unconscious of the fetid odor Cancer or lupus affecting the nasal chambers or encroaching on them may also be responsible for a discharge possessing an unpleasant smell

Cerebrospinal Rhinorrhea This is a rare condition caused usually by operative or accidental trauma especially fractures at the base of the skull It may begin without any known cause In instances of increased intracranial pressure spinal fluid may escape to the nose and in the presence of the accompanying disturbances of the sensorium and the recumbent position of the patient it drains back into the throat and causes gurgling coughing and choking Ordinarily there is a constant drip of the clear fluid from the nose

The diagnosis depends on the presence of a clear fluid from the nose which does not contain mucin or albumin but contains sugar as evidenced by the reduction of copper in a copper containing solution such as Fehling s solution

Epistaxis A discharge of blood from the nose may be a result of either local or general causes It is usually a capillary oozing and on inspection the bleeding area is seen to be congested ecchymotic or superficially ulcerated A common site of the bleeding spot is on the cartilaginous septum Less frequently it arises from the posterior end of the middle turbinated body or in children with adenoids from the vault of the pharynx or at times from the accessory sinuses Specular inspection under a good light clots having been removed by swab or spray is necessary to de termine the exact location of the hemorrhagic point If epistaxis occurs during sleep the blood may run into the pharynx and after having been swallowed may be vomited or if clinging to the pharynx be hawked up thus simulating hematemesis or hemoptysis Nosebleed from local causes is usually unilateral but if the nosebleed is due to blood changes or general diseases it is likely to take place from both sides

Ordinarily epistaxis is not sufficiently severe to produce constitutional symp toms but after trauma or operations on the nose or in cases of hemophilia it may be profuse or continue long enough to threaten life seriously although an actual fatal result is extremely rare

NASAL CAUSES OF EPISTAXIS Aside from traumatic causes caries or necrosis of the nasal bones ulceration from a foreign body or other causes polyp new growths varicosities or chronic nasal catarrh may be responsible for epistaxis Hypertension with accompanying arteriosclerosis and alcoholism may render the vessels liable to rupture The latter causes rarely produce nosebleed

GENERAL CAUSES OF EPISTAXIS People of full habit may have a rather frequently recurring nosebleed Delicate children suffer from it especially at puberty and after

vascular nature of the spiders may be demonstrated by pressing the point of a pencil on one of the arteries where it is seen to emerge from the deep layers of the skin During application of pressure in this manner the vessel is obliterated

Acute redness of the nose with pain and swelling especially of one ala may be the result of a small pustule or boil

In a young person a superficial ulceration of the wing of the nose usually painless beginning as a reddish papule spreading in various directions and healing in one portion while breaking down in another is probably a tuberculous ulcer In an elderly patient a small hard scabbing ulcer somewhat painful gradually extending and perhaps attended with involvement of lymph nodes is an epithelioma

Collapse of the Alae Nasi The alae nasi collapse when an unnatural weakness of the lateral aspects of the lateral walls of the distal part of the nose is present This condition is a developmental defect often inheritable The nose is long and narrow in contour The collapse of the nares occurs during inspiration and thus produces varying degrees of nasal obstruction

Atresia of the Alae Nasi In congenital atresia of the alae nasi there is a failure of perforation or absorption of epithelium of the developing nose Acquired atresia of the alae may be engendered by healing tuberculosis syphilis or trauma

Acting Nares Regurgitation of Fluids In some sensitive and neurotic individuals the nostrils dilate with each inspiration especially under mental excitement Aside from this acting nares indicate dyspnea or anoxia of various origins and among other conditions are very noticeable in emphysema asthma pneumonia obstructive diseases of the larynx and the broken compensation of cardiac valvular lesions

If on the patient s attempting to swallow fluids a portion escapes through the nostrils one of three conditions is usually present cleft palate paralysis of the soft palate (diphtheritic) or bulbar paralysis

Pain in or Around the Nose Burning or smarting sensations usually attend coryza or other acute catarrhal inflammation If great pain in the nasal chambers is present it may be a symptom of an ulcerating lesion or of an impacted foreign body Severe pain just above the root of the nose accompanied with fever may be due to inflammation of the frontal sinuses Pain in the nose and cheek is due to inflammation of the antrum Pain referred to the nasal cavities and ears is due to inflammation of the eustachian tubes Inflammation of the eustachian tubes is often associated with diseases of the nasal passages A sensation of dryness is a common complaint in the early and late stages of coryza and is a persistent and annoying symptom of atrophic rhinitis

Inflammation and engorgement of the turbinates ostia nasofrontal ducts and superior nasal spaces are responsible for most of the pain from most of the paranasal structures The pain emanating from the nasal and the paranasal structures is relieved by shrinkage of the nasal structures

Diseases of the superior nasal structures cause headaches primarily in the front and top of the head and between the eyes Diseases of the middle and inferior nasal structures cause headaches over the zygomas and temples and in the teeth and jaws

When the turbinates are painful the pain is augmented by shaking the head and by lowering it between the knees The presence of purulent secretion in the sinuses is not necessarily the basis for headache

Local vasomotor changes in the erectile tissues of the nose occur as accompaniments of stress exhaustion sexual excitement and various emotional states which may or may not be associated with symptoms If there should be inflammation of the nose these changes enter awareness and thus cause symptoms Furthermore congestion of the nasal mucous membranes may occur without inflammation as an accompaniment of anxiety or resentment

To test the function of the olfactory nerve nonirritating substances must be used as pungent vapors affect mainly the trigeminal nerve Suitable odorous agents are musk oil of cloves and peppermint in convenient containers It should be remembered that the perception of any single odor is lost in 3 or 4 minutes but is regained after 1 minute of rest It is essential that the odorous material should enter the nostrils as a vapor or in a state of fine division the act of smelling is usually assisted by sniffing which is a modified inspiration It is also necessary that the mucous membrane should be moist in order for the odorous particles to enter into solution on its surface

The clinical symptoms relating to the sense of smell are *anosmia* loss of the sense of smell *hyperosmia* increased sensitiveness of the sense of smell and *parosmia* subjective perversion of the olfactory sense

Anosmia The loss of the sense of smell in the majority of instances is due to local disease of the nasal mucous membrane namely chronic rhinitis particularly the atrophic variety and polyps or other new growths The inhalation of irritating vapors or extremely foul odors may temporarily or permanently abolish the sense of smell Paralysis of the trigeminal nerve will impair the olfactory power of the affected side Loss of the power of smell may be a symptom purely neurotic of neurasthenia and hysteria

Frequently the sense of smell is abolished because of injury or lesion of the olfactory bulb or tract Falls or blows on the head affecting the nerve in its course may produce as the only symptom of injury a persistent anosmia Caries of the bones supporting the tract or bulb and basal meningitis or tumors involving the nerve may be instrumental in causing olfactory anesthesia Possibly from atrophy of the nerves anosmia may be one of the symptoms of locomotor ataxia Anosmia is some times congenital resulting from imperfect development of the olfactory nerve tissue Partial anosmia may be due to a lesion of the uncinate gyrus or to disease of one hemisphere

Hyperosmia Hyperosmia or abnormal sensitiveness of the sense of smell is in its slighter degrees not an uncommon symptom in neurotic individuals More rarely the sensitiveness is extreme and is symptomatic of hysteria and neurasthenia

Parosmia Olfactory sensations without a physical basis the apparent odors usually being unpleasant or offensive (*kakosmia*) are not infrequent as hallucinations in the psychoses A bad odor may constitute the premonitory aura of epilepsy Subjective kakosmia has been associated in a limited number of cases with tumor of the hippocampus

Olfactory Acuity and Appetite Elsberg and Levy introduced a simple method of measuring olfactory acuity Measured volumes of odorous air were injected into both nostrils The smallest volume which produced a sensation of odor was taken as a measure of the olfactory threshold Using bottles containing constant amounts of ground coffee as sources of odorous air Goetzel and associates measured the diurnal variations in olfactory acuity They found that on days when lunch was omitted the olfactory acuity increased gradually from the early morning until the end of the work day This increased olfactory sense was accompanied with a parallel increase in the hunger sense This basic pattern was modified on days when lunch was eaten during the midday rest period As a result of the lunch the olfactory acuity fell precipitously to a relative olfactory insensitiveness as the sensation of hunger was converted into one of satiety This dullness of the olfactory sense persisted for about 3 hours when a rapid increase in olfactory acuity set in accompanied with a return of the hunger sense

Postnasal Drip A postnasal drip is of importance mainly from the local dis comfort and anxiety that it gives the patient who is thus affected

Postnasal drip is produced by many and varied etiologic agents It is often associated with the various forms of rhinitis lymphoid hyperplasia of the pharynx and enlarged adenoids and lingual tonsils It is caused by irritations and by dust on the nasal mucous membranes as well as by excessive tobacco smoking and medica tions in the way of nasal sprays and nasal washes containing drying agents The

exertion under a hot sun It is a common complaint of mountain climbers at considerable altitudes and inhalation of very hot or very cold air may induce it In suppressed menstruation it may be vicarious The chronic anemias are frequently attended by epistaxis It is a symptom of leukemia purpura haemorrhagica and scurvy and is the most ordinary form of hemorrhage in hemophilia Less commonly it may be the result of cardiac hypertrophy and valvular disease enlarged bronchial lymph nodes pleurisy with large effusion emphysema and the strain of pertussis

Epistaxis is a symptom of diagnostic value in the prodromal stage of typhoid fever and is an occasional event in other infections such as erysipelas pyemias malarial fever viral rickettsial and leptospiral infections

HEREDITARY HEMORRHAGIC TELANGIECTASIA (Osler s Disease) This telangiectasia is a rare hereditary disease characterized by the presence of multiple acquired angiomas of varying distribution and number with a tendency to bleed spontaneously or from slight trauma The disease has been traced through six generations in one family In about 20 per cent of the cases there is no family history of a similar disease

The disease is regarded by some as simple ectasia of preexisting vascular channels By others it is regarded as multiple acquired neoplastic angiomas arising from endothelial rests of embryonal origin This is suggested by the following observations Epistaxis usually predates the development of visible mucosal or cutaneous lesions satellite lesions develop in the region adjoining the site formerly occupied by angiomas destroyed by treatment the lesions have a universal distribution and some are of great size they have a tendency to disappear spontaneously while new angiomas appear

The first symptom is abnormally profuse epistaxis beginning at about puberty This is followed by the development of multiple telangiectasias of the skin and mucous mebranes from the age of 25 to 35 years All symptoms tend to reach their greatest severity from 30 to 39 years of age

Fissure on the Nasal Vestibule A fissure on the nasal vestibule is a common complication or sequence of a nasal discharge from any cause It often happens during the course of coryza and hay fever A less common cause is syphilis The fissure is painful and the tip of the nose is tender The skin about the fissure is reddened

Sneezing This is a spasmodic expiration caused usually by direct rarely by reflex irritation of the sensory nerves of the nose which occurs as an early symptom of coryza measles pertussis asthma and hay fever Small doses of the iodides in susceptible individuals and large doses in many others will induce it as well as the inhalation of various irritants such as dust pepper and snuff Prolonged paroxysms of sneezing have been asserted to be of hysterical origin Syringing or manipulation of the external auditory canal will sometimes provoke a reflex sneeze A common cause of sneezing is a hypersensitive lining of the nose such as is present in vasomotor rhinitis

Picking at the Nose Picking at the nose with the finger by a child was formerly considered to be a sign that the child had intestinal worms It has been said that picking at the nose is common in those who have brain tumors However picking at the nose is an expression of nervous tension and shyness Nervous or shy persons may have in due time either worms or brain tumors but picking at the nose is a sign of neither intestinal parasites nor brain tumors

The Sense of Smell The olfactory bulbs and their associated parts are in reality portions of the brain The peripheral nerves arising from the bulbs are distributed to the mucous membrane of the upper portion of the nasal septum the superior and middle turbinates and the proximal part of the nasal septum These distributions of the olfactory nerve can be identified by their yellowish brown color The free end of an olfactory cell is rodlike and reaches the surface of the epithelium where it divides into small projecting olfactory hairs of microscopic dimensions The center for the sense of smell is said to be in the uncinate convolution

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complaint is frequent hawking and clearing the throat especially marked on arising in the morning and during or soon after meals The patient often interprets the symptoms as being due to sinusitis Cough and hoarseness may be the chief complaint when laryngitis is present Often there are sensations of tightness of dryness and fullness of the throat There may be difficulty in nasal breathing Some impairment of hearing may result from obstructions of the eustachian tubes Often in the morning in attempts to clear the throat and the back of the nose of the tenacious secretion vomiting may ensue Generally speaking a postnasal drip does not produce disease of the lungs or of the stomach swallowing of the material is harmless and normal

Rhinitis *Acute rhinitis* is characterized successively by a feeling of dryness in the nose nasal obstruction anosmia and then a watery irritating nasal discharge Accompanying these manifestations often are sneezing lacrimation headache fullness and discomfort in the ears and fever

Acute rhinitis accompanies many and varied conditions It is often the beginning of an infection of the upper part of the respiratory passages

Chronic Rhinitis Chronic rhinitis often follows an acute attack of rhinitis or it may result from a succession of mild attacks of coryza or may be left after measles scarlet fever or other specific infection This condition is not contagious It is frequently met with in those exposed to dusty occupations The commonest symptom of chronic rhinitis is nasal obstruction which seldom is complete It is generally intermittent being worse after meals in certain conditions of weather and at night During the day it may be negligible but at night the nostrils become so obstructed that mouth breathing with all its discomforts is present On examination there is an engorgement of the vessels with chronic inflammatory changes in the mucosa

Hypertrophic Rhinitis The complaint in hypertrophic rhinitis is of abundant sometimes thick mucopurulent discharge occasionally stained with blood if much force has been used in expelling it There is nasal obstruction worse at night than by day and influenced by gravity so that the nasal chamber toward the pillow is most obstructed The obstruction may be sufficient to induce mouth breathing The sense of smell is diminished hence much of the perception often attributed to taste is lost The voice loses its nasal resonance Discomfort or actual pain may be caused by pressure of a congested turbinal body on the septum

Atrophic Rhinitis Atrophic rhinitis is a chronic rhinitis of unknown origin and occasionally occurs in several members of a family The affection tends to disappear after middle life The condition is much less common in men than in women It is characterized by atrophic changes in the nasal mucosa and turbinated bones abnormal patency of the nasal fossa and a mucopurulent discharge with foul odor The sufferer however is rarely aware of the odor for as a rule the disease causes a loss of the sense of smell The discharge tends to dry and to crust and the separation of the crusts from the nose may cause slight bleeding

Atrophic rhinitis symptomatically may resemble the symptoms of foreign bodies rhinolith empyema lupus or suppurating adenoids

Vasomotor Rhinitis (*Spasmodic Rhinorrhea*) Vasomotor rhinitis may be defined as an intermittent engorgement of the nasal mucous membrane associated with a free discharge of dilute mucus frequently with paroxysmal sneezing and suffusion of the eyes Often there are present a hypersensitivity of the nasal mucosa and some external irritation acting on the nasal mucosa The psychoneurotic element is an important causative factor Examination of the mucous membrane during an attack shows passive engorgement

The attacks come with great suddenness often in the early morning and commence with a feeling of tickling or dryness in the nose Perhaps the same sensation in the conjunctivae and a sudden attack of violent sneezing usher in a discharge of profuse watery mucus from the nostril The sneezing may be so violent and uncon

trollable as to cause ecchymosis of the conjunctivae Often as soon as the mucous membrane has been wetted by secretions the attack ceases

Nasal Leprosy Leprosy of the nose is manifested by chronic nasal obstruction bloody nasal discharge and marked crusting As time passes there is destruction of parts of the nose with perforation of the nasal septum and maximal external nasal deformities

The diagnosis is established by the presence of other symptoms of leprosy and by the demonstration of the bacilli in the nasal discharges Often the diagnosis of leprosy can be established by the administration of 10 drops of a saturated solution of potassium iodide in water to increase the nasal secretions At the height of the secretion about 2 hours after administration of the drug the nasal septum and the nasopharynx are swabbed with a cotton applicator and the material is fixed on a glass slide by heat stained with carbolfuchsin and counterstained with methylene blue *Mycobacterium leprae* is acid fast

Screwworm Infestation (Cochliomyia americana) of the Nose The common site of screwworm infestation in man is the nose but infestation of nasal sinuses pharynx throat mouth ear orbit eyeball and open wounds may also take place The chief predisposing factor in rhinal myiasis is a pre-existing pathologic condition of the nose A cut scratch or abrasion of the skin is necessary to attract the female screwworm fly Sleeping in the open invites infestation The eggs hatch out 24 hours after being deposited in a favorable environment The symptoms include partial obstruction to breathing on the affected side and a feeling of discomfort accompanied with a strange sensation at the root of the nose that becomes an intense pain Sneezing may occur at the onset and severe headache may persist A serosanguineous nasal discharge of offensive odor accompanies these symptoms With the onset of this discharge the patient may clear his throat repeatedly expectorate frequently and cough up a purulent material that may contain living or dead larvae Removal of the larvae is accomplished mechanically and by the use of drugs Chloroform applied locally and as a vapor is the drug of choice Although screwworm infestation occurs chiefly in the Southern states cases have been observed as far north as Missouri and Illinois

Diseases of the Nasal Septum The septum in most noses is somewhat irregular and rarely is placed evenly between the nares

Ulcers A commonly occurring ulcer of the nasal septum develops as the result of habitual picking of the nose with the index finger a habit caused by nervous tension The inception is manifested as a septal erosion from repeated trauma The urge to do more picking of the nose is made stronger by the irritation and discomfort The septum may be deeply injured and the mucosa may be denuded

Rare causes for ulcers of the nasal septum are syphilis and trauma from irritating chemicals and particulate matter such as organic and inorganic particles or dusts

Once the ulcer is established there ensue nasal irritation and epistaxis If syphilis is the cause there is a foul nasal discharge with destruction of the septum

The ulcer is identified by endoscopic examination

Deformities The deformities are classified as spurs deviations and combinations of spurs and deviations If of a sufficient degree these deformities originate the complaint of nasal obstruction and inability to clear one or both nostrils Any of the sequelae of mouth breathing however may be due to aberrations of the nasal septum

A spur is found to consist of a crust a spina or a thickening It is chiefly cartilage but it is often present at the junction of cartilage and bone both of which enter into its formation Deviations of the nasal septum are almost always more pronounced in the cartilaginous portion of the septum than in the bony portion

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of the nose is determined by roentgenologic examination The diagnosis of malignant disease is established by biopsy

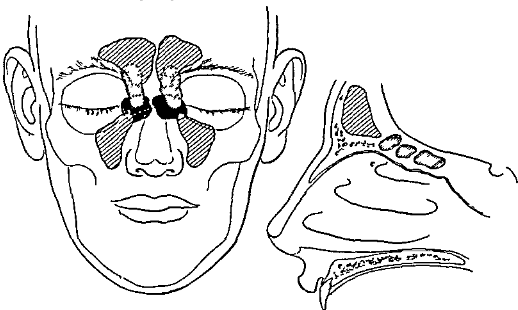


Fig 4 2 Accessory nasal sinuses

The Accessory Sinuses The accessory sinuses or air cells of the nose are the frontal ethmoidal sphenoidal and maxillary (Fig 4 2) They vary in size and form in different individuals These sinuses are lined with ciliated mucous membrane which is directly continuous with that of the nasal cavities

The *frontal* sinuses are frequently the seat of suppurative inflammation which gives rise to pain and tenderness in the supra orbital region and to a discharge from the cor responding nostril This discharge can be seen coming from beneath the anterior extremity of the middle turbinated bone Owing to the proximity of the opening into the maxillary sinus pus coming down the hiatus from the frontal sinus may pass into the maxillary sinus thus simulating disease or producing disease of that cavity too

The *ethmoidal* sinuses or cells anterior middle and posterior three on each side lie between the sphenoidal sinus posteriorly and the lower extremity of the frontal sinus anteriorly The region of the ethmoidal cells is that from which mucous polyps of the nose take their origin These polyps are a common accompaniment of suppuration of the accessory nasal cavities Caries affecting the anterior cell of each sinus may extend into the orbit and the pus may form a fluctuating tumor above the inner canthus of the eye It is to be recalled that a meningocele may be present in this same location

The *sphenoidal* sinuses are the most posterior lying still farther back than the eth moidal sinuses They open into the spheno ethmoidal recess above and posterior to the superior turbinated bone Discharge from them goes into the pharynx and can be seen with the rhinoscopic mirror

The *maxillary* sinuses lie one on each side of the nose beneath the orbit and to the outer side of the nasal fossa The sinus opens into the nose by a slit like opening into the middle meatus about its center posterior to the hiatus semilunaris and 1 inch (2 5 cm) above the floor of the nose When the opening is close to the hiatus liquid may run into it from the hiatus

The inflammatory and infectious diseases of the maxillary sinus originate by exten sion from either the nose or the teeth The sinus is the seat of tumors often malignant and of inflammation the latter accompanied by an accumulation of mucus or pus The walls of the sinus are thin so that a tumor bulging forward causes a protrusion of the cheek The tumor presses inward and obstructs the breathing through the side of the nose or it pushes upward and causes protrusion of the eye by encroaching on the orbit

Perforation Perforating ulcerations of the bony part of the nasal septum are often due to syphilis but when the cartilaginous septum only is attacked, the ulceration is probably not due to a specific process An ulcer of the nasal septum may form and perforation may take place without the patient knowing of its occurrence The symptoms complained of are a sense of dryness irritation in the nose the separation of crusts and occasionally free epistaxis When perforation first occurs there is often a whistling sound caused by the passage of air through the small opening This gives much annoyance to some patients but they can be reassured since the sound generally ceases when the opening increases to a larger permanent position

Nasal Obstruction One of the most frequent and most important consequences of nasal disease is obstruction to respiration The harmfulness of mouth breathing entailed in cases of severe disease is definite

Since nasal obstruction is a factor in numerous nasal affections and since many of its possible consequences are not always recognized an enumeration of the causes and symptoms is worth while Inactive alae nasi which permit of diminished ventilation of the nose and arrested development of the nasal passages lead to *deviation of the nasal septum and often to defective development and growth of the* upper maxillae with arching of the palate V shaped alveolar arch and crowded teeth There is a tendency to frequent and prolonged nasal catarrh and later to hypertrophic rhinitis Postnasal catarrh and increase in adenoid tissue often follow

Catarrh of the eustachian tubes in either acute or chronic otitis media may develop as a part of the general pathologic processes that follow nasal obstruction

The symptoms of nasal obstruction are difficulty in clearing the nose of mucus noisy nasal respiration with sniffling heavy breathing and snoring loss of nasal resonance alterations of the voice anosmia or loss of the sense of smell and deformity of the thoracic wall Well developed mouth breathing is associated with a typical facies dry mouth spongy gums imperfect mastication of the food and slowness in eating or necessity of bolting the food

Nerves of the Nose In addition to the olfactory nerve the nose is supplied by the nasal infratrochlear and infra orbital branches of the fifth nerve hence the eyes water when the nose is injured In certain cases of neuralgia affecting the ophthalmic division of the fifth nerve pain is felt along the side of the nose Because the nasal nerve enters the skull from the orbit through the anterior ethmoidal foramen it may be involved in disease of the ethmoidal sinuses

Nasal Polyp Polyps are the commonest tumors of the nose They are benign tumors which usually do not become malignant however they tend to recur after removal

The causes of nasal polyps are unknown These tumors are often associated with hyperplastic rhinitis

Mucous polyps arise from the outer wall and principally from the margin of the middle meatus and the cells of the ethmoidal labyrinth The symptoms of nasal polyp develop slowly and the patient sometimes becomes so gradually accustomed to the condition that the disease is often well marked when the patient is first examined The symptoms are those of nasal obstruction and its consequences

Nasopharyngeal Polyp Nasopharyngeal polyps are usually unilateral and solitary A single polyp in the nasopharynx may have its origin in *the antrum and* by growth pass into the nose through an accessory ostium maxillare The symptoms are those caused by one sided nasal obstruction and catarrh There is increasing discomfort as the polyp descends into the pharynx

Malignant Tumors Malignant tumors of the nose are rare The symptoms of a malignant nasal tumor may commence by the presence of nasal obstruction and discharge and bleeding spontaneously or on slight manipulation Spontaneous bleeding occurring in a middle aged patient arouses suspicion of the presence of malignant disease The extent to which a malignant tumor has involved the nose or the bones

Inflammation and obstruction may extend to the lacrimal passages the con junctival and finally the orbital tissues producing orbital cellulitis and periostitis Extension of infections from an accessory sinus may cause a great swelling and ptosis of the eyelids chronic conjunctivitis blepharitis photophobia dilatation of the pupil and blepharospasm In such ocular involvements exophthalmos and some impairment of the mobility of the eyes are common These conditions are serious illnesses and always are accompanied by marked constitutional symptoms which are manifested by high fever and violent pain in this region

The aural symptoms arising from a chronic suppuration of the sinuses are tinnitus vertigo and earache Noises in the ear as a result of eustachian catarrh may be the first discomfort to attract the patient s attention

Headache face ache hemicrania and neuralgia though commonly of origins other than the sinuses often require an exploration of the sinuses Pain in the face arising from sinus disease may present a morning periodicity increasing for some hours after the patient arises in the morning and often disappearing as the day goes on This periodicity has been explained as the result of accumulation and retention of secretion during the night The secretion then escapes from the cavity during the first hours of physical activity

The diagnosis of chronic sinusitis is made on the basis of the symptoms and of examination for signs of rhinitis particularly in the region of the ostium of the sinus together with evidences of discharge in the nose and nasopharynx The pres ence of polyps at once points to chronic ethmoiditis In many cases roentgenologic examination will reveal changes in the bone or thickening of the linings of the cavities of the sinuses Roentgenograms of the sinuses often reveal the presence also of hyperplastic thickening of the membranes and of formation of polyps in the sinuses Finally an exploratory puncture and washing of the sinuses to reveal pus if present may be required for diagnosis

There are but few physicians left who regard a chronic sinusitis as of much importance as a focus of infection The prognosis is usually for spontaneous recovery or at least for temporary alleviation of symptoms

Voice Dynamics and the Ethmosphenoid Epiglottidean Syndrome The nose and the nasal cavities act as resonating chambers for the voice whereby sounds pro duced in the larynx are amplified and changed in timbre A lack of nasal resonance as observed in nasal obstruction is termed rhinolalia clausa

The patients complain usually in the morning that they are not in good voice The alteration in voice and the hoarseness clear up during the day Vocalization and swallowing usually dispose of the effete mass that has accumulated during the night The patient finds it difficult to start phonation and his throat tires quickly

In some patients the symptoms may be referable to involvement of the muscles of the pharynx and their nerve supply To a singer this may be a devastating malady Pressure in the laryngeal structure from even a small amount of pus may be suffi cient to interfere with the normal emission of the pus by coughing As the disorder advances pus accumulates in large quantities and remains in evidence when the epiglottis is examined

In some patients there may be a partial or total loss of taste or smell a ticklish and tight feeling about the throat and blocking of the ears with or without spasmodic cough Less frequently patients hear tingling noises an echo or a vibration Pus may be found in the nasopharynx and redness of the tympanic membrane without an impairment of hearing

Cysts and Benign Tumors of the Sinuses The maxillary sinuses may contain mucous retention serous periodontal or dentigerous cysts Mucous retention cysts are the common cysts often reported by the roentgenologists A cyst of the peri odontal type arises from the root of a pulpless tooth The bone is eroded from the

Diseases of the Accessory Sinuses Suppuration may arise primarily from direct infection of an accessory sinus or secondarily from some intranasal disease Among the acute infectious diseases which give rise to sinusitis are influenza pneumonia measles scarlatina smallpox cerebrospinal meningitis diphtheria erysipelas and more rarely mumps and gonorrhea Coryza and all processes of nasal disease associated with pus formation may induce empyema in the accessory cavities The anterior group of sinuses is much more frequently affected than are the posterior groups Of the anterior cells the maxillary sinuses are the ones most often involved the ethmoidal next and the frontal sinuses least often

Acute Sinusitis This forms a part of many acute catarrhs which invade the sinuses by direct extension from nasal coryza Acute exacerbations are not uncommon in the course of chronic empyema Bathing in a public swimming pool particularly if diving is done may lead to infection of the nose and sinuses Acute sinusitis is generally met with in adult life but is not unknown in childhood

The symptoms of an acute sinusitis frequently form a part of acute nasal coryza However if during an acute nasal coryza there occurs pain in the region of a sinus accompanied by tenderness on pressure over the cavity sinusitis is suspected Lacrimation photophobia edema or a light congestion of the eyelid with proptosis and deep seated headache also suggest the implication of a sinus More than one cavity may be affected at the same time The occurrence of a chill or of a rise in temperature would indicate more than a simple catarrh in a patient suffering from acute rhinitis Relief of pain follows a discharge of mucus which may be bright yellow and blood stained or mucopurulent or relief follows a free gush of pus which sometimes is very offensive both to smell and to taste

On examination there is usually evidence of acute rhinitis or the middle turbinal will be congested infiltrated and pushed against the septum and the meatus below it will be clogged with stringy mucus and later on with mucopurulent material Transillumination may direct attention to the maxillary sinus and puffiness of the forehead or fullness or duskiness of the eyelid on one side will point to the frontal or the ethmoidal cavities When the posterior group is affected the rhinoscopic mirror will show a deeply congested appearance of the pharynx and anterior sphenoidal wall and mucopus will ultimately be visible above the superior turbinal on the posterior pharyngeal wall Most patients who have acute sinusitis recover spontaneously Complications very rarely result but sometimes occur during the course of a chronic sinusitis as the result of an acute exacerbation

Chronic Sinusitis Chronic sinusitis in the accessory sinuses is common In many cases roentgenologic examination reveals clouded mucous membrane of the sinuses but this does not necessarily imply active disease

The maxillary sinuses are those most often affected Patients afflicted with empyema or chronic sinusitis frequently seek relief on account of discharge or of obstruction or of a chronic cold in the head

The symptoms of chronic sinusitis may be grouped as (1) nasal symptoms (2) nasopharyngeal symptoms (3) ocular symptoms (4) aural symptoms and (5) headaches

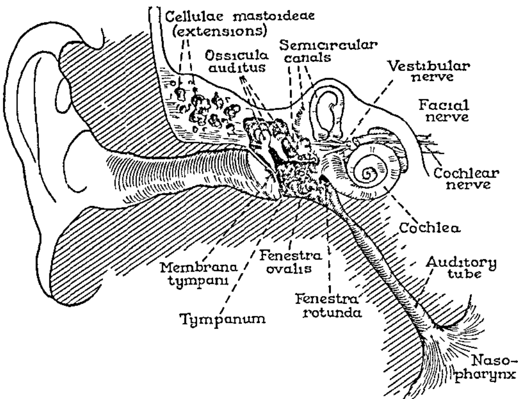
The nasal symptoms are nasal obstruction and nasal discharges The obstruction may be unilateral or bilateral It is usually more marked in the morning hours than at other periods and is more intense with the increase of nasal inflammation The discharges may flow posteriorly and the obstruction may be slight and the secretion scant in amount so that the patient for years has swallowed it unconsciously Disorders of the senses of smell and taste are often indicative of the disease There may be parosmia or anosmia or there may be kakosmia The presence of kakosmia is suggestive of sinus suppuration The patient is seldom aware of the odor

The symptoms in the nasopharynx and the larynx are those of a postnasal irritation or low grade inflammation

and frostbite is therefore common Injuries and wounds of the cartilage occur readily and are slow to heal

Discoloration A livid or *bluish* tint of the external ear is usually indicative of general cyanosis A frostbitten ear is at first white but when it becomes thawed the skin is cyanotic A *blue black* discoloration of the cartilages of the ear occurs in ochronosis (usually with alkaptonuria) A swollen discolored pinna may be due to *injury* or *hematoma*

Angioma or enlargement of the blood vessels not infrequently affects the external ear and not only may disfigure but also may show a tendency to extension to the scalp face and neck In some instances these angiomas are jet black and fissured and contain hair



Redrawn from M [illegible] published drawings by permission of W B Saunders Company

Fig 4 3 The ear

The External Meatus The external auditory meatus extends from the concha to the drum head (Fig 4 3) In order to look into the ear and see the membrane it is necessary to straighten the canal by pulling the auricle outward upward and backward The general direction of the canal in the adult person is from without downward inward and slightly forward In a child upward traction is not so necessary as in the adult In childhood the length of the canal is approximately the same as in adult life but the bony part is still in a cartilaginous condition

The external opening of the meatus is oval but farther in the canal is more nearly circular In examining the meatus the otoscopic attachment of an ophthalmoscope or a speculum and head mirror are used

Below and in front of the ear is the temporomaxillary joint and just posterior is the glenoid lobe of the parotid gland When the gland is inflamed and swollen it presses on the cartilaginous canal and produces pain and in case of suppuration

tooth to the sinus by means of granulation tissue formation Dentigerous cysts are of congenital origin and are mentioned in association with tumors of dental origin

These cysts do not produce symptoms unless they have attained space occupying proportions sufficient to give pressure symptoms The symptoms consist of pain There may be swelling of the cheek and bulging of the lateral part of the adjacent nasal cavity Diagnosis is established by roentgenographic examinations and needle puncture Small cysts require no treatment

Papillomas adenomas lipomas hemangiomas osteomas chondromas gliomas and mixed tumors occur mainly in the frontal and ethmoidal sinuses These tumors usually do not give symptoms until they have occupied all the available space in the sinus After this they produce symptoms from pressure and by eroding adjacent bone and interfering with the function of other organs such as causing a unilateral exophthalmos by extending into the orbit

Malignant Tumors of the Sinuses Malignant tumors which affect either the paranasal sinuses or the nasal cavities as a rule arise in the sinuses These tumors consisting equally of carcinomas and sarcomas arise most frequently in the maxillary sinuses However they do arise in the ethmoidal and the frontal sinuses

When the tumor extends from the sinus it erodes bone There is pain and often there are severe epistaxis and nasal obstruction A purulent bloody nasal discharge is common Malignant tumors in the antrum cause pain in the teeth and cheeks sensory disturbances of the skin and swelling of the cheek There is often bulging into the nose and hard palate or unilateral exophthalmos The lymph nodes on the ipsilateral side are enlarged and metastasis ensues to distant regions

The diagnosis is made on the basis of roentgenographic studies which reveal a mass or defect in the bones of the affected sinus Biopsy of tissue from the sinus or from the nose is conclusive

Malignant Tumors of the Nasopharynx Malignant tumors of the nasopharynx are usually highly anaplastic carcinomas usually designated by such terms as lympho epithelioma or transitional cell carcinoma They metastasize early and it is usually the metastatic lesions that first attract attention—swelling in the neck or pain or deafness in one ear Headache sore throat epistaxis and nasal obstruction are frequent complaints Cranial nerves may become involved with resultant diplopia or optic atrophy Signs due to involvement of the fifth seventh and other cranial nerves are occasionally demonstrable The local lesion in nasopharyngeal lympho epithelioma may escape detection by expert otolaryngologists for as long as 6 months after the tumor has been histologically identified in a metastatic cervical node

THE EAR

The external ear (auricle pinna) is composed mainly of a cartilaginous framework covered with thin skin The lobe which is the lower part of the ear is composed of dense connective tissue containing fat The large concavity leading into the meatus is the concha

The external auditory meatus the tympanum and the eustachian tube are formed from the first branchial cleft in the fetus A failure of any portion of the cleft to close normally may leave small sinuses or depressions in the neighborhood of the ear and these may persist after birth as congenital fistulas More severe deformities may result from cleft abnormalities resulting in absences of parts or all of the ear

Ears that are unusually prominent long or misplaced with absence of helix antihelix or lobule are marks of imperfect physical development These abnormalities do not necessarily carry stigmas of degeneration An unusual protrusion of large auricles from the sides of the head is the commonest of all ear deformities

The loss of the auricle does not cause much if any decrease in sound perception The subcutaneous tissue of the ear is well supplied with blood Owing to the absence of fat, the exposed condition of the blood vessels renders the ear sensitive to cold

Neuralgia or recurring pain in one ear similar to the episodes of pain from tic douloureux is experienced Arthritis of the temporomaxillary articulation is accompanied by earache Angina pectoris and if in the right ear aneurysm of the innominate artery may give earache Affections of the tongue may be accompanied by otalgia Herpes zoster oticus may be preceded or followed by severe pain in the ear

Discharges From the Ear A fracture of the base of the skull may cause an aural hemorrhage followed by the escape of *clear cerebrospinal fluid*

Rupture of the tympanic membrane either as a result of injury or consequent on otitis media may be responsible for a discharge of *blood*

A flow of *pus* otorrhea may be either acute or chronic and while it may be due to a polyp or abscess of the external meatus it is in the majority of cases an evidence of otitis media with or without complication

Diseases of the Membrana Tympani Reflected light from a head mirror or an otoscope directed through the meatus by a speculum reveals the membrana tympani Extending downward from its center is a small cone of light Any depression or bulging of the membrane will cause this cone of light to be altered in its position or not be present at all From the center of the membrane upward extends a line which indicates the point of attachment of the long handle of the malleus Stretching across the upper portion is the membrana flaccida

During the course of acute rhinitis pharyngitis or influenza blebs or bullae filled with blood serum or blood may develop in the skin of the deep portion of the external canal and on the drum These may be present before there are symptoms of otitis media The blebs appear as purple blisters which break at touch and yield bloody serum leaving the skin or the drum wet and red Soon the blebs break and cause a thin bloody discharge Often synchronously with the sanguineous discharge from the ear there is relief of the pain

Rupture The ear drum may be ruptured by the introduction into the canal of pins or sticks for scratching or removal of wax flat handed slaps or boxes on the ear the compression of explosion as by artillery fire too rapid decompression as occurs in divers and in occupants of aircraft after rapid descent from high altitudes

The symptoms of ruptured drum are sudden and intense pain in the ear followed by deafness loud tinnitus and perhaps vertigo nausea and vomiting If infection follows and acute otitis develops otoscopic examination reveals fresh or dry blood in the canal and on the drum The perforation may be visible Bloody crusts in and about the ruptured membrane will be present for several days Increasing pain fever and a seropurulent discharge denote the presence of otitis media

Diseases of the Middle Ear ***Otitis Media*** (*Infections of the Middle Ear*) *Infections of the middle ear pass up the eustachian tube from the pharynx and the* nasal cavities The infection may pass thence to the mastoid antrum and mastoid cells On pus formation and suppuration of the middle ear the pus usually obtains an exit by perforating the tympanic membrane and discharging through the external auditory meatus

In instances of retropharyngeal abscess or suppuration of the middle ear pus may pass down the eustachian tube and escape through the nose

In a retropharyngeal abscess originating in a vertebra the pus may pass through the fascia or behind it extend laterally and appear under the sternocleidomastoid muscle While behind the fascia the pus may be in contact with the base of the skull and if so the infection may pass through the porous bone and involve the meninges and the brain In rare instances the pus may perforate the bone below and anteriorly and involve the jugular vein and the internal carotid artery

In suppurations of the middle ear if the posterior bony wall of the auricular canal is perforated the infection may involve the facial nerve with facial paralysis ensuing The brain also may be reached by the infection extending through the internal ear through the fenestra ovalis and rotunda and the internal meatus and

pus may discharge through the external meatus gaining access to the canal through fissures in the cartilage The cartilaginous portion of the meatus contains sweat glands sebaceous glands and hair follicles There are only a few glands in the upper posterior portion of the bony meatus

Impaction of Cerumen Cerumen is the normal secretion of special ceruminous glands in the cartilaginous portion of the external auditory canal Excessive accumulations of cerumen may give symptoms When symptoms are present the mass has formed and it may then move with motion of the head or may swell on the entrance of sweat or water and produce complete obstruction A complete obstruction causes deafness tinnitus and a full stopped up feeling in the ear At times there is a cough from a vagus nerve stimulation There may be pain or vertigo if the mass presses against the drum If the auditory canal has been filled with the wax there may be dilatation of the canal or erosion of the skin and granulation tissue may result Bleeding ensues on removal of the wax healing follows readily

Furuncle A furuncle of the external auditory canal is due either to the entrance of pyogenic organisms into the subcutaneous tissue by way of hair roots or follicles or to an infection of sweat glands

The commonest predisposing cause of furuncle of the auditory canal is an acute or a chronic dermatitis of the canal The process begins with pain of increasing intensity in the ear with throbbing there is pain on movement of the lower jaw and wide opening of the mouth may be impossible because of the pain Local pressure on the pinna or auricle or motion of this part is painful There may be a general aching and at times a fever

The diagnosis of furuncle of the auditory canal is made easily on inspection of the canal especially if the ear drum is not congested or is not perforated A roentgenologic examination should be made if there is doubt about involvement of the mastoid cells

External Otitis Some forms of external otitis may be due to otomycosis An otomycosis is usually an accompaniment of a mycotic infection of the skin The ear feels sore and tender chewing may be painful The auditory canal often contains soft semifluid wax and debris of flecks of a white foamy substance As the disease progresses the ear is tender and chewing is painful The canal is sometimes completely filled with soft moist sebaceous like detritus which often has a greenish color due to a secondary infection After the ear has been cleansed the canal wall is found to be red with excoriation of the epithelial lining The drum is not affected In the severely affected ear the canal is swollen often obliterated and very painful The pain is worse at night than by day and there may be fever An otomycosis is diagnosed by identifying the fungus in the discharges from the affected ears

The external auditory canal is affected in many forms of dermatosis

Pain in the Ear Earache Usual Causes The most frequent cause of severe pain in the ear (earache) is otitis media secondary to many illnesses such as scarlet fever epidemic influenza acute simple rhinopharyngitis or tonsillitis measles or diphtheria It is commonest in children and may explain an obscure infantile illness with fever

Other Causes Pain referred to the ear or about the mastoid process when there is no focal cause for such symptoms is a reflex or referred otalgia Common causes of these secondary otalgias are exposed dentine and inflamed pulp or dying nerves of the teeth on the same side particularly in the mandibula unerupted impacted or infected molar and wisdom teeth which may cause severe deep boring earache of long duration Acute inflammation or ulcerative processes exposing nerve ends in the nasopharynx pharynx laryngopharynx and larynx may give rise to otalgia Malignant growths even small ones of the nasopharynx are prone to cause this symptom A sphenopalatine ganglion neuralgia often causes pain in the ear or in the region of the emissary vein on the posterior surface of the mastoid process

persistent mucopurulent discharge constantly or intermittently In such a condition healing may eventually take place but there is permanent impairment of hearing Owing to impairment of function of an injured labyrinth an impairment of hearing may become manifest after all signs of infection have ceased

The serious complications of acute otitis media are suppurative mastoiditis serous or purulent labyrinthitis meningitis extradural brain abscess and lateral sinus thrombosis

Suppurative Mastoiditis Suppurative mastoiditis is a complication of otitis media or it occasionally occurs after fractures of the skull Once the infection is established in the mastoid cells it tends to persist and to suppurate

The common etiologic agent is the hemolytic streptococcus However any one of the pyogenic organisms may be present such as the staphylococcus or the pneumococcus

There is a rapid vascular engorgement with edema and infiltration and the production of a mucopurulent exudate The mucosal cells degenerate so that the bone is deprived of blood supply and the cell walls disintegrate Large cavities are present in the well pneumatized mastoid form and if the cortex is thin or thinned perforation ensues If the aditus and antrum are large enough to furnish adequate drainage into the middle ear resolution can take place

The symptoms depend on the severity of the suppurative process Pain is present in the ear and over the mastoid area Fever (temperature up to 103 F or more) which is present for several days may then subside except for afternoon rises The recurrence and the continuation of the fever indicate a more severe process There are nausea vomiting anorexia constipation and increasing weakness

Tenderness which may be widespread is present over the mastoid antrum There is often edema of the skin and soft parts and swelling of the posterosuperior wall of the external auditory canal The tympanic membrane is bulging and perforated There is often deafness of the conduction type without tinnitus

A polymorphonuclear leukocytosis is the rule The urine may contain albumin depending on the amount of fever The roentgenogram reveals the degree of bone changes as well as the degree of pneumatization of the cells The latter is helpful in determining prognosis for the highly pneumatized cells are more easily and widely destroyed

Labyrinthitis The labyrinth is often affected from the toxic agents of infections without the entrance of bacteria Bacteria may enter the labyrinth as the result of trauma surgical or otherwise or from middle ear disease In widespread labyrinthitis there may be destruction of essential structures and loss of function

The invasion of the cochlear portion of the labyrinth causes tinnitus and deafness Invasion of the vestibular apparatus produces varying degrees of vertigo nystagmus temporal or retro ocular headache vomiting and mild weakness of the facial muscles In diffuse involvement of the labyrinth the patient becomes very ill with severe vertigo which makes it impossible to stand alone The patient lies absolutely still The room seems to turn toward the diseased ear The slightest movement brings on violent vomiting

The diagnosis of labyrinthitis depends on the accurate testing and evaluation of the turning caloric and fistula tests by one trained in employing these tests and interpreting the findings Acutely ill patients are not subjected to these tests

Tuberculosis Tuberculosis of the middle ear is rare except in young adults during the advanced stages of pulmonary tuberculosis The infection seems to be brought to the ear as a blood borne infection or when there are serious deep lesions of the pharynx or nasopharynx In the latter instances the infection reaches the ear through the eustachian tube

Once in the ear a tuberculous infection causes widespread destruction of the tympanic membrane [illegible] ne of the labyrinth In

then to the brain The infection may extend to the attic and the antrum and perforate the roof of these structures and form a subdural abscess in the middle cerebral fossa Extension of an inflammation from the antrum and mastoid cells may enter the posterior cerebral fossa to involve the lateral sinus causing a lateral *sinus thrombosis* or produce a cerebellar abscess

ACUTE OTITIS MEDIA In an acute otitis media the fluid content of the middle ear is either serous or purulent Mild infections of the middle ear occur which do not perforate the tympanic membrane When the fluid of the middle ear is serous the otitis may be of allergic origin However all forms of otitis media are to be considered infectious in origin until proved different The bacteria most commonly involved are the hemolytic streptococci the *Hemophilus influenzae* and all forms of pneumococci staphylococci and in rare instances such organisms as the *Klebsiella pneumoniae* In most cases these organisms enter the auditory canal through ethmoid cells or through the eustachian tube by continuity of tissue aided by such mechanical devices as blowing of the nose sneezing and coughing In instances in which the drum has been perforated previously the organisms may enter from the external auditory canal However in most instances of acute inflammations of the middle ear the organisms enter from the nasal cavities the paranasal sinuses the nasopharynx and the pharynx

In the beginning of otitis media the pain may be mild but as the inflammatory process spreads the pain becomes more severe The pain is often the boring lancinating type that spreads into the occiput temples and teeth and is made worse by coughing sneezing or swallowing Except in very mild infections there is fever The height of the fever indicates to some extent the severity of the infection In mild cases the temperature will reach 101 or 102 F (38 3 or 38 9 C) in severe infection it may be as high as 104 F (40 C) The fever usually continues 4 or 5 days with morning remissions and ceases by lysis Synchronous with the onset of the fever there are malaise anorexia vomiting pain in the back and limbs and constipation The pain and often the fever disappears on perforation of the drum

The drum is usually retracted It may appear injected or may have a grayish color or a yellow cast and often is shiny At times bubbles of fluid or a fluid meniscus may be visible through the tympanic membrane

CHRONIC SUPPURATIVE OTITIS MEDIA Chronic suppurative otitis media is manifested by an intermittent purulent discharge from the ear which accompanies respiratory infections During acute exacerbations the discharge from the ear increases During an exacerbation if the discharge stops often there are pain and fever

The diagnosis is established by otoscopic examination The perforation of the drum is identified A central perforation without involvement of the annulus tympanicus differentiates this form of otitis media from the more severe forms

The perforation of the tympanic membrane in the severe necrotic types of otitis involves the peripheral parts of the drum The opening is irregular in shape and may represent destruction of from one half to two thirds of the drum There may be granulation tissue or a polyp inside the perforation

Masses of debris appearing on the attic and the tympanic atrium which arise from squamous epithelium growing through the perforation in the drum are known as *cholesteatoma* The term is derived from the content of fat and cholesterol of the masses of debris

The perforation may be a large defect in the tympanic membrane through which the promontory of the ossicles may be visible Deafness of the conduction type is variable in its severity Roentgenologic examination may reveal a nonpneumatized type of mastoid process

COMPLICATIONS OF OTITIS MEDIA The complications of otitis media depend on the severity of the disease The infectious process may become subacute with a

vertigo of central nervous system origin develops slowly over a long period There may be associated nystagmus and falling The diagnosis of these lesions however lies within the field of neurology and neurotology

Types of Dizziness and Vertigo DeWeese emphasized that it is not wise to focus all of the attention on hearing and labyrinthine function for in the great majority of patients complaining of dizziness the results of functional examination of the ear will be normal

Cerebral anoxemia may produce dizziness Arteriosclerosis and hypertensive cardiovascular disease often produce a constant feeling of uncertainty or intermittent mild attacks of dizziness In postural hypotension dizziness occurs when the patient rises suddenly from a recumbent or sitting position to the standing position Pernicious anemia or any severe anemia may produce transient or recurrent dizziness as a result of cerebral anoxemia

Sudden and more prolonged cerebral anoxemias are produced by paroxysmal auricular fibrillation aortic stenosis with insufficiency arteriosclerotic heart disease (Adams Stokes disease) and carotid sinus hypersensitivity In some cases of dizziness from these causes it may be necessary to withhold a final opinion until the patient can be observed during an attack

Dizziness is not a feature of brain tumor or other destructive disease inside the calvarium unless there is depression of function of the inner ear on one or both sides Dizziness is a feature of lesions of the cerebellopontine angle and of the brain stem In cerebellopontine angle lesions there is loss of hearing tinnitus corneal anesthesia fifth and seventh cranial nerve involvement ataxia and intracranial hypertension

Chronic and recurring dizziness cannot be attributed to head injuries unless there is a persistent loss of hearing in a patient who had normal hearing prior to the accident However concussion can give rise to dizziness tinnitus headache and other symptoms without any positive physical findings perhaps as the result of multiple small hemorrhages Compensation neurosis and outright malingering are common and may cause diagnostic difficulties in those who have had head injuries

In labyrinthine hydrops there are paroxysmal attacks of whirling vertigo usually with abrupt onset almost always accompanied with nausea and vomiting These attacks last an hour or two and subside There is complete freedom from vertigo between attacks There is an accompanying perceptive hearing loss frequently fluctuating almost always progressive and usually more severe in one ear than in the other Tinnitus most commonly persistent between attacks and frequently fluctuating is present The associated hearing loss is often a flat low tone perceptive loss and the caloric reaction is hypoactive

Acute toxic labyrinthitis often follows an acute febrile disease indiscretion in consumption of food or alcohol or the use of any type of drug In this syndrome there is a gradually increasing whirling vertigo lasting 1 to 3 days followed by a slow subsidence of symptoms within a period of a week There is no associated hearing loss or tinnitus

When the end organ is suddenly and completely destroyed either by injury or by hemorrhage there are a sudden overwhelming vertigo nausea vomiting loud tinnitus and complete loss of hearing A slow compensation takes place over a period of 10 to 20 days and gradually the opposite labyrinth controls reasonably well the function previously presided over by both labyrinths

The commonest cause of dizziness is a functional disorder The diagnosis should be made on the basis of positive evidences of the existing neurosis This is one of the most important diagnoses to be made lest there be created an iatrogenic disorder

Meniere s Disease Meniere s disease is characterized by sudden attacks of vertigo with tinnitus or incomplete deafness in one or both ears

Meniere s disease may be attributable to disease of the labyrinth either hemor

vasion of the facial canal and paralysis of the facial nerve ensue A destructive form of labyrinthitis with deafness may occur These widespread destructions may take place with a minimum of symptoms

Examination discloses that the drum is perforated and discharges a foul purulent material There is a perception type of deafness The diagnosis is established by the demonstration of tubercle bacilli on culture and by guinea pig inoculation of the suspected material

Syphilis Congenital syphilis frequently engenders deafness by destruction of the cochlea and semicircular canal apparatus Infants thus affected are born with total deafness In some instances of congenital syphilis osteomyelitis of the labyrinth or syphilitic neuritis of the cochlear nerve does not occur until between the fifth and fifteenth years of life

The diagnosis is based on the history and the presence of Hutchinson s triad (1) interstitial keratitis (2) Hutchinson s teeth and (3) deafness Often perforation of the bony part of the nasal septum is present

In acute acquired tertiary syphilis which is present in the nasopharynx the process will extend to the membrana tympani Generally such an extension is made by pyogenic organisms rather than by the *Treponema pallidum* Once in the ear the syphilitic process is very destructive The labyrinth is destroyed and sequestra are formed There is a profuse purulent discharge from the middle ear There are severe labyrinthine symptoms which soon subside as total deafness ensues The diagnosis is established from the history the examination and the strongly positive serologic reactions

Tinnitus Subjective sounds heard in the ear are tinnitus aurium Sounds which are vaguely situated in the head are tinnitus cerebri Sounds of either type are usually heard on one side only and are described variously by the patient

Tinnitus is a common symptom and may be due to so many conditions that in a given case it is often impossible to determine its cause Tinnitus is a response of the auditory part of the eighth cranial nerve to abnormal stimuli

It is helpful to bear in mind that the most frequent causes of tinnitus usually ringing in the ear as described by the patient are psychoneurosis with anxiety and consequent vasomotor irregularity diseases of the ear such as impacted cerumen otitis with fluid in the middle ear and obstruction of the eustachian tube blood diseases as in anemias and senescence An attack of migraine is frequently preceded by or accompanied with sudden loud noises

Tinnitus with deafness may be due to the administration of quinine as well as salicylic acid and its compounds Ergot in considerable doses may give rise to ring ing in the ears because of its probable action in producing cerebral anemia Various subjective sounds due to toxemia circulatory disturbances or aural disease may be present in some of the *infections* particularly cholera malaria and rickettsial fevers

Tinnitus or ringing in the ear is a symptom of ear disease in 8 out of 10 patients affected with otologic disorder When tinnitus is associated with an otologic disease there is always an accompanying deafness which may be slight low pitched roaring or rumbling constant or throbbing or high pitched whistling or ringing

Unilateral tinnitus and very severe or total deafness of the perception and nerve type with or without vomiting and vertigo followed by numbness in the face diplopia occipital frontal pain stiffness of the posterior neck muscles unsteady gait and ataxia choked optic nerve head loss of corneal reflex weakness of the lower part of the face of the same side with a dead labyrinth on a caloric test and Kernig s sign are highly suggestive of an expanding intracranial tumor or if there is fever a brain abscess

Vertigo True vertigo may be caused by labyrinthine disease lesions of the central nervous system such as acoustic tumor and cerebellar disease Often the

ing which follows inflammation of the middle ear resulting from simple or specific infectious rhinopharyngitis postnasal adenoids enlarged faucial tonsils nasal polyps and nasal stenosis from any cause Wax in the meatus aural polyps parotitis or other inflammations and growths may produce deafness by closing the external meatus A unilateral deafness of sudden onset is often of central origin During the course of meningococcic meningitis or meningoencephalitis typhoid fever influenza scarlet fever or measles deafness may occur which follows the destruction of the nerves the nerve ganglia or the structures of the labyrinth by the meningeal inflammation

With advancing age there is a loss of hearing for high pitched tones deafness of the nerve type with tinnitus in one or both ears

Overuse or hypersensitiveness to quinine and salicylates may cause a degeneration of the nerve cells The use of these drugs during pregnancy may affect the unborn child through the placental circulation In lead poisoning the lead may act on the vessels of the cochlea and on the brain Phosphorus carbon monoxide chenopodium arsenic mercury morphine and aniline dyes have produced nerve deafness Streptomycin at times seems to have a specific transitory toxic effect on the cochlea and vestibular branches of the auditory nerve The deafness and tinnitus caused by streptomycin usually disappear after its discontinuance However the vestibular effects of the drug often are permanent

Permanent deafness may begin abruptly on the fourth and fifth days of epidemic parotitis (mumps)

Hemorrhage into the labyrinth if both divisions of the eighth nerve are involved may produce sudden but permanent deafness and loss of function of the semicircular canal apparatus since it destroys the essential end-organs

Traumatic and *occupational deafness* may occur as a result of different forms of acoustic trauma as a blast injury or prolonged exposure to loud noises In hysteria and malingering mere pretense of deafness is common

The distinction between ordinary middle ear deafness and nerve deafness is made by testing with a watch or a vibrating tuning fork If the deafness is due to *aural disease* the ticking of a watch or the vibrations of the tuning fork are heard faintly or not at all when the instrument is held at varying distances from the ear these noises become distinctly audible when the watch or the handle of the tuning fork is placed in contact with the skull or the mastoid process (bone conduction) The sound is heard best by the nonaffected ear if deafness is due to disease of the auditory passages by the affected ear if deafness is due to obstruction of the air passages In *nerve deafness* the watch and the fork are heard indistinctly or not at all either in contact or at a distance In ordinary deafness the nerve is normal and can appreciate vibrations brought by the bone but through some fault in the mechanism aerial vibrations are not transmitted to the nerve endings In nerve deafness the nerve is at fault and cannot appreciate vibrations no matter how well they may be conducted

The *Weber test* determines whether nonaural impairment is of obstructive or nervous origin by comparing the bone conduction of the two ears by means of a tuning fork

The *Rinne test* is used to compare the duration of bone conduction with that of air conduction for the ear being tested The patient is Rinne positive when air conduction is greater than bone conduction The patient is Rinne negative when bone conduction is greater than air conduction

Otosclerosis In otosclerosis there is the formation of new soft bone in the capsule of the labyrinth which replaces the normal bone with ensuing progressive deafness due to fixation of the footplate of the stapes

Otosclerosis is a distinct nosologic entity due to a genetic mutation with dominant characters In the heterozygous state otosclerosis rarely becomes manifest if it does it is less severe than in the homozygous state and appears only at advanced age This genetic factor may not be manifest for several generations but despite this histologic examination of the organ of hearing reveals the existence of otosclerotic foci

The clinical manifestations of heterozygous otosclerosis can be influenced by

rhagic or degenerative to hydrops of the labyrinth to suppurative or other disease of the middle ear to a vasomotor disturbance of the vessels of the semicircular canals which may accompany a neurosis and to disease of the centers of hearing and equilibration

The vertigo of Meniere s disease is sudden in its onset and the patient may fall before there is time to prevent it There may be a transitory loss of consciousness The face is pale and covered with perspiration Nausea and vomiting ensue There is present a tinnitus which may consist of hissing buzzing roaring or throbbing noises The attacks tend to recur several times in a day or may be months apart Rare symptoms during the attack are diplopia and nystagmus double vision or oscillatory movements of the eyeballs The deafness usually is progressive but in complete affecting one or both ears In intervals between the attacks there may be few symptoms but a continuous giddiness is a common complaint

In those who have symptoms of Ménière s disease a test of the reaction of the vestibular function and the testing of hearing are necessary The determination of the function of the vestibular portion of the end organ as it functions through the semicircular canals is obtained through the Barany test (caloric test)

The value of the caloric test (Barany s symptom) depends on stimulating the endolymph in the semicircular canals to move On sudden movement of the endolymph in a normal individual nystagmus and vertigo ensue The stimulus is effected by turning the subject or by instilling hot or cold water into the external auditory canal in order to change the temperature of the endolymph

In carrying out the turning test the subject is turned in a special chair with head in proper position at a certain rate of speed and for a definite number of times The caloric test is conducted by instilling cold water (68 F 20 C) or hot water (112 F 44 4 C) into the external auditory canal By either the turning or the caloric procedure nystagmus and vertigo of definite types and duration will be produced in normal per sons Absence of reactions or change in character of reactions has pathologic significance Interpretations of the abnormal reactions must be made by those experienced in conduct ing the test This test should not be used when the ear drum is perforated

Diagnosis depends on the history and the positive findings as recorded and interpreted

Motion Sickness The essential cause of motion sickness for example seasick ness train sickness or air sickness is the abnormal stimulation of the vestibular apparatus by the unusual motion to which the patient is subjected Psychic influ ences play some part in the production of motion sickness Stimulation of the semi circular canals is especially likely to cause symptoms this occurs for example dur ing pitching or rolling of a ship or when an airplane suddenly drops in a down draft The symptoms are vertigo nausea and continuing vomiting and sometimes diarrhea with intense headache and mental depression In severe attacks the vomiting and diarrhea may produce profound dehydration and the lack of food intake may cause acidosis so that shock ensues

Deafness The hearing mechanism consists of two portions the conductor and the receptor The external ear and the middle ear represent the conductor the in ternal ear represents the receptor

More instances of deafness result from advancing years than from disease of the tympanic membrane the middle ear or the eustachian tube Disease of the audi tory nerve its nucleus or its cortical center gives origin to nerve deafness Often no cause at all can be found to account for deafness Deafness may be congenital or acquired

Congenital deafness may result from defects in development of the hearing apparatus as the result of intra uterine syphilis or German measles or from the ad ministration of quinine to the mother prior to the birth of the child Congenital deafness often coexists with deaf mutism

Acquired deafness usually results from loss or impairment of the power of hear

The color of the eye is not a simple hereditary component For instance of blue eyes alone there are 9 shades or classes which are transmitted in heredity in no less than 45 possible combinations

The Palpebral Fissure the Eyelashes and Eyebrows The breadth or the transverse diameter between the openings of the eyelids is the palpebral fissure The palpebral fissure varies racially as follows narrowest in the Mongolian peoples widest in the whites and intermediate in the Negroes The height of the upper lid is classed as low mid and high This measurement decreases with age owing to growth of the facial skeleton and development of the supra-orbital complex

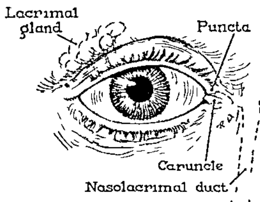


Fig 4 4 Region of the eye

The eyelashes and the eyebrows vary greatly among individuals in appearance and color They are not always the same color as the hair In men the color of the beard corresponds with the color of the hair except that red beards may occur when the hair is dark Eyelashes and particularly eyebrows add much to the physiognomic effect of the face In women the eyelashes and eyebrows are so altered cosmetically that descriptions of their natural appearance are nullified Women tweezer off or paint on eyebrows to suit their particular type of beauty They may glue on false eyelashes

With the advent of middle age a man may be made aware of his years by the barber saying Shall I trim the eyebrows sir? In other words a brief description of beginning old age in men is the time when hair begins to grow where it should not be and ceases to grow where it should be

Headache of Ocular Origin Unilateral head pain of ocular origin is common Pain radiating through one eye may be due to a ciliary spasm localized neuralgia or the spasm of a single ocular muscle A shooting knifelike intermittent stabbing pain when due to ciliary spasm or a spasm of a single muscle is associated with the use of the eyes Localized viral disease along nerve trunks may cause prolonged local pain Postneuritic pain may be severe Inflammation of the gasserian ganglion and irritation of the internal carotid artery from aneurysm are uncommon causes of eye pain Glaucoma is often the source of unilateral or bilateral eye pain

Nearsighted persons do not have headache or head pain unless the nearsightedness is unequal or severe or unless they are abusing their eyes Abuse in the use of nearsighted eyes may produce severe headache nausea and vomiting

Farsighted persons are likely to have frontal headaches which are moderate to severe and are present almost daily in the afternoon or evening Farsightedness sometimes is definitely associated with certain types of work

If there is an inequality in the amount of error in the two eyes the pain may be severer over one eye and commoner as a cause of headache

Errors in refraction usually produce frontal vertical or bitemporal pain which may be associated with slight dizziness Nausea is usually relieved by going to bed or resting or taking a single tablet of acetylsalicylic acid It is made worse by sewing playing cards and other close application An error of refraction associated with a muscle error is almost certain to produce symptoms of headache such as pain or nervousness irritability exhaustion nausea and loss of weight The ophthalmologist is uncertain about a patient s muscle balance if the patient has some constitutional

exogenous factors and also by endogenous milieu These factors especially the endogenous milieu will precipitate the manifestations at an earlier age even in those who have been conditioned for this sort of deafness

The manifestations consist of a progressive loss of hearing for low tones by air conduction with or without tinnitus There is an apparent improvement of the hearing when exposed to noise (paracusis willisiana) However the loss of hearing does not vary from day to day except for its progression toward deafness Eventually there is deafness of a mixed form

The findings on examination depend on the progress of the disease There is present conduction deafness in both ears without a previous history or findings suggestive of otitic suppuration in a patient of less than 30 years of age The genealogic history reveals that a similar form of deafness may have been present in previous generations In all the deafness has progressed more rapidly during gestation or serious illnesses than at other periods Owing to nerve degeneration there is finally an impairment of hearing of the mixed form

Interpretation of Hearing Tests Guild has expressed the opinion that nothing is gained clinically by making hearing tests which cannot be interpreted with reasonable certainty in terms of causative lesions In other words if there be no histologic evidence to support or to refute the diagnostic interpretations customarily made of the data obtained by most of the elaborate time-consuming technics in the interpretation of these technics only a description of the kinds of hearing defects found by the tests made and not a diagnosis is given Guild has expressed an opinion that for most patients a few simple tests of hearing afford all the information the clinical otologist needs to supplement or to confirm the impressions gained from the history and the physical examination and has reached the conclusion that a clear distinction should be made between tests for research purposes and tests for clinical purposes

The histologic findings in instances of conduction deafness of similar degree as given by Guild lend emphasis to the statements that the practical conclusion is that despite any advances in methods of testing hearing reliance on the clinical history and the physical examination must continue in order to determine why a patient is hard of hearing and what can or should be done about it

THE EYE

The signs and symptoms referable to the eye are of importance not only from the standpoint of disability from eye disease but also with reference to disorders of the nervous system and in connection with diseases of other organs

The Color of the Eye The pigment of the eye is contained in the conjunctiva the sclera the iris the choroid the ciliary processes and the retina The color of the eye ranges from brown through gray and green to blue The lack of pigmentation of the eye in albinism is abnormal but not pathologic The impairment of vision produced by albinism is mainly due to photophobia and nystagmus

There are three major types of distribution of the pigment of the eye the mixed the rayed and the zoned In the eye of *mixed* colors the periphery of the iris is of some light clear shade specks or dots or perhaps strands of different color sometimes variations of the color of the iris but commonly brown yellow or orange encircle the pupil In the *rayed* eye the pigmentation varies in intensity and radiates from the pupil somewhat in the manner of spokes from the hub of a wheel In the *zoned* eye the pigmented part of the iris is a well defined band around the pupil the periphery is gray green blue or yellowish

In the newborn the eyes are blue or violet As a child grows the area around the pupil becomes pigmented so that a brown areola and a light background are characteristic Among European peoples the color of the eyes darkens during adolescence In general the coloring of the eyes tends to keep pace with darkening of the hair In the aged the coloring of the eyes decreases and the eyes appear faded

Ectropion Ectropion is an eversion of the eyelid with exposure of the conjunctival surface of the lids it may affect both lids The condition is secondary to scarring of the lids However it does occur in the aged as a part of the relaxation of the skin and the orbicularis oculi muscle Atrophy may occur with ectropion as a part of an affection of the facial nerves causing paralysis of the orbicularis oculi muscle and under these circumstances the ectropion is known as the paralytic atrophic type This affects only the lower lid

Trichiasis Trichiasis is an inversion or ingrowing of eyelashes so that they rub against the cornea This condition is manifested by congestion of the cornea pain lacrimation photophobia opacities vascularization and ulceration The most common cause of trichiasis is chronic trachoma

Swelling Puffiness The eyelids particularly the lower are swollen in general anasarca and are often the earliest seat of edema of renal origin Anemia and hypoproteinemia may be attended by swelling of the lids A bloated appearance of the eye possibly with ecchymoses is common in severe cough and pertussis Angioneurotic edema and phenomena indicative of the allergic state may be seen in this locality Edema of the lids and forehead which occasionally occurs in neurotic and adolescent individuals is often of allergic origin

Because of the anatomic connection between the blood vessels of the face and those of the cranial cavity cerebral thrombosis may give rise to a swelling of one or both eyelids with some protrusion of the eyeball the puffiness subsequently extending to the face Likewise infections of the face may extend through these same vessels and produce thrombosis of the cerebral venous sinuses

Erysipelas and the stings of insects may be the cause of swelling so great as to prevent opening of the eye Severe coryza hay fever arsenic poisoning and iodism may induce noticeable swelling of the lids So also the lids are swollen in measles variola varicella and rickettsial infections to mention only a few causes for swollen lids

Hordeolum (Sty) A small painful abscess at the edge of the eyelids is a sty or hordeolum When sties occur in successive series it may be indicative of the over use of defective eyes Small single and superficial sties are of purely local origin A sty is to be differentiated from chalazion (cyst of a meibomian gland) a slow growing tumor which may become inflamed and suppurate

A cystic swelling of some duration toward the inner canthus is a *mucocele* (chronic dacryocystitis) which may become inflamed and suppurate After measles scarlet fever ophthalmia or other diseases in which there is conjunctival inflammation there may be an acute inflammation of the lacrimal sac

Ankyloblepharon Ankyloblepharon is an adhesion of the margins of the upper and lower lids It may be either a congenital condition or due to burns It may be partial or complete and is often combined with symblepharon as a part of widespread scarring and deformities of the eyelids

Blepharophimosis Blepharophimosis is a contraction of the palpebral fissure at the outer canthus The outer angle is obscured by a vertical fold of skin from contraction following prolonged epiphora and blepharospasm

Blepharospasm Blepharospasm is characterized by an increased winking of the eyes Essential blepharospasm is the normal amount of winking of the eyes both lids moving synchronously Symptomatic blepharospasm may accompany disease of the eye as of the trigeminus nerve

The term blepharospasm also may designate a slight narrowing of the palpebral aperture which may be of congenital origin or form ankyloblepharon

Lagophthalmos Lagophthalmos is a condition of incomplete closure of the palpebral aperture when the eyes are shut It may be due to narrowing of the lids from contraction of a scar or to congenital deformity ectropion paralysis or loss of power of the orbicularis oculi facial paralysis or proptosis due to orbital tumor or

disease such as hyperthyroidism hypothyroidism, paralysis agitans, *encephalitis or* diabetes

There is a definite seasonal and periodic increase in head pain originating in the ocular muscles Such headaches become more frequent in October reach a peak in January and fall off in the spring These headaches may be related to inactivity for when the days become longer and outdoor exercise is resumed the incidence decreases

The child who has a nervous breakdown or who is inattentive the adult who has a headache at noon which is relieved by lunch and then has a recurrence at about 3 or 4 P M the student who cannot concentrate and the convalescent patient who has headache may belong to the group of persons who have faulty poor or inadequate ocular musculature It is common to observe instability of eye musculature in patients undergoing a period of physical debility Many of these patients who have ocular muscle imbalance are unaware of their difficulty

Those who have hyperthyroidism hypothyroidism and other glandular disturbances producing headaches often have been given a pair of glasses during the acute stage of the general disease When recovery was attained the tone of the ocular muscles improved and then the glasses were not suitable for them

THE EYELIDS

The skin of the lids is thin and the subcutaneous tissue loose and devoid of fat For these reasons after injury blood finds its way readily into the lids and shows plainly beneath the skin as in the familiar black eye

Mongoloid Fold The mongoloid fold occurs commonly among Mongolians and occasionally among the whites

The epicanthal fold among the Chinese for instance is on the whole much thicker than among white peoples This condition of thickening in the Chinese eyelid is due to a characteristic disposition of the lower part of the orbicularis oculi muscle plate whereby as seen in sagittal sections the lower marginal fibers are disposed backward in a curve or even a kink

Coloration of Skin of the Eyelids A darkening or *duskiness* of the lids and under the eye is seen in some women and men Duskiness in either men or women may be prominent in the anemia and pallor of brunet persons or after fatigue mental excitement loss of sleep or severe pain and in exhausting diseases Dark circles around or under the eyes are often hereditary and have no other significance This condition is chiefly an affection of brunets who have thin fair skin

One or more soft and very slightly elevated yellow patches seen on the upper eyelids of middle aged women or of elderly men or women but most frequently in the soft tissue below the lower lids are manifestations of xanthelasma These spots or plaques varying in size from 1 mm to 1 cm in diameter when on the eyelid are situated near the inner angle they may be either unilateral or bilateral They may extend to or primarily involve the skin under the lower lid These are of no diagnostic significance

Coloboma of the lid is a triangular notching of the upper lid at the junction of its inner and middle thirds Damage may be done to the eye if the deformity is not corrected

Tophi As in the ear, so may small nodules of sodium urate significant of gout form in the eyelids

Entropion Entropion is a rolling in of the margins of the eyelids and eyelashes It is observed when there are scars or spasms of the lids Cicatrical entropion most commonly affects the upper lid Spastic entropion which is due to spasm of the palpebral portion of the orbicularis oculi muscle almost always affects the lower lid These conditions are manifested by mechanical irritation and injury to the cornea from the turned in eyelashes There are congestion pain lacrimation and photophobia If not corrected opacity, vascularization and ulceration of the cornea ensue

Carcinoma of the lid is usually a basal cell epithelioma which is termed rodent ulcer Malignant tumors of the eyelids occur in older people and more frequently on the margin of the lower than of the upper lid

The tumor begins as a small elevation in the epithelium which ulcerates early and appears as an ulcer with indurated edges The ulcer slowly spreads *Highly* malignant squamous cell epithelioma occasionally occurs in young persons and spreads rapidly to the adjacent lymph nodes along the sides of the neck

Sarcomas are rarely present on eyelids

THE LACRIMAL APPARATUS

The lacrimal gland consists of two portions an orbital or superior portion and a palpebral or inferior portion The lacrimal gland opens by several fine ducts into the fornix of the conjunctiva

The lacrimal canal embracing the sac and the nasolacrimal duct each about 12 mm in length extends from just above the internal tarsal ligament or tendo oculi to the inferior meatus of the nose Below the palpebral ligament the sac is comparatively weak and here it is that distention occurs and pus makes its exit

Dryness and Moisture of the Eye The eye may become *dry* and *glazed* in prolonged fevers as well as in disease attended by lagophthalmos

Lacrimation an increased secretion of watery fluid usually accompanies irritation or inflammation of the conjunctiva and accordingly is present in measles influenza pertussis and rickettsial infections in the early stages in hay fever asthma coryza trifacial neuralgia facial paralysis and iodism and when a foreign body is present in the eye The tears may overflow the edge of the lids (epiphora) because of displacement of the puncta lacrimalia or obstruction of the duct Cold winds and irritating vapors stimulate the flow of tears

Hyposecretion of lacrimal fluid is manifested by a feeling of dryness and burning smarting sensation in the eyes photophobia and impairment of the vision Hyposecretion or deficiency in lacrimal secretion does result from extirpation of the lacrimal gland but is usually caused from the scarring of the conjunctiva that follows trachoma and xerophthalmia

Stagnation of the contents of the lacrimal sac is quickly followed by infection and inflammation and is termed dacryocystitis The lacrimal sac may become obstructed by either congenital causes or diseases of prenatal influences or as a result of chronic nasal infection

Reduced production of lacrimal fluid is found in a condition known as keratoconjunctivitis sicca often associated with defective secretion of the salivary gland and arthritis In vitamin A deficiency dry greasy areas known as Bitot s spots may develop on the bulbar conjunctiva in the interpalpebral space

Epiphora is an overflow of tears and is a symptom in affections of the tear conducting apparatus The common causes of epiphora are exposure to wind or smoke affections of the nose and irritation of the retina by bright light It may also be dependent on increased secretion which occurs in facial paralysis or from affections of the ophthalmic branch of the trigeminus nerve due to foreign bodies and inflammation of the eye Epiphora is commonest in old people especially those who are exposed to the open air in cold weather and often is without manifest lesions of the conjunctiva or tear passages

The ophthalmologist investigates the lacrimal passages by digital or instrumental pressure over the lacrimal sac and by injection of physiologic saline solution through the inferior punctum with the lacrimal syringe If on pressure over the lacrimal sac a watery viscid mucopurulent discharge escapes through the punctum it indicates an obstruction of the nasolacrimal duct When obstruction is present it may be situated in the nasolacrimal duct or in the lacrimal sac or at the junction of the common canaliculus with the lacrimal sac

to exopthalmic goiter In the last named disease lagophthalmos is conjoined with protrusion of the eyeball (proptosis) and inability to follow with the upper lid a downward movement of the eyeball (von Graefe s sign) Lagophthalmos may be due to laxity of the tissues and absence of reflex blinking in patients who are extremely ill or moribund Incomplete closure of the lids during sleep is commonly present in exhausting diseases As the eyeballs are normally turned upward in sleeping the scleras are thus exposed giving a ghastly expression to the face Owing to exposure the cornea becomes epidermoid (xerosis corneae) or keratosis sets in

A similar rolling up of the eyes and imperfect closing of the lids are frequently seen in hysteria in various convulsive disorders during the convulsion and immediately after death See ptosis (fall) of the upper lid page 120

Blepharitis Blepharitis is characterized by reddened thick inflamed and crusted edges of the eyelids The edge of the lids may possibly be affected by minute ulcers or pustules The condition often results from an allergy a previous ophthalmia or an attack of measles or it may accompany anemia Often however the cause is unknown

Blepharitis is often the source of a chronic conjunctivitis The etiology of the conjunctivitis may be suggested by the appearance of the blepharitis For instance a mixed bacterial infection is often irregular in distribution and omits the meibomian glands and follicles of the cilia whereas the staphylococcal variety involves these structures

Residual swelling from recurrent sties indicates a residual staphylococcal infection of the lid margins which may be the source of a chronic conjunctivitis

Meibomianitis is often present in conjunctivitis Expression of the contents of the meibomian glands and observation of the expressed material are regularly a part of the examination of the edges of the eyelids when disease is suspected An inflammatory enlargement of one of the meibomian glands is termed *chalazion*

Swelling redness and dilatation of lacrimal puncta are characteristic of streptomycotic concretions which constitute an important cause of unilateral conjunctivitis The diagnosis can be made by expressing material from the punctum and detecting the streptomyces on microscopic examination or on cultures

Molluscum contagiosum is a contagious disease caused by a filtrable virus The incubation period is variable from 14 to 50 days It is transmitted by direct contact Man is the only known host The virus is cosmopolitan in distribution and affects all ages The disease is characterized by the formation of firm rounded pearly white skin nodules or tubercles varying in size from one to several millimeters in diameter The lesions appear on the eyelids the face the scalp arms legs buttocks mucous membranes of the mouth and genitalia as single or multiple nodules The soles and the palms are never affected These nodules usually present a central depression and contain semifluid caseous matter or solid matter made up of fat epidermis and peculiar capsulated bodies the molluscum corpuscles

Chronic edema of the eyelids due to blockage of lymph vessels is a characteristic feature of *lymphogranuloma inguinale keratoconjunctivitis* and can be considered almost pathognomonic of the infection

Herpes zoster ophthalmicus is an involvement of the eyelid when there is herpes zoster of the ophthalmic division of the fifth nerve The lid is affected by the characteristic lesions of herpes zoster The pain is severe The lesions heal leaving scars which may be disfiguring

Tumors *Papillomas* of the eyelids are common along the edges of the lids They should be removed Papillomas also include the verruca or wart Xanthelasma naevus hemangioma and lymphangioma may be present on the eyelids

Small cysts having transparent contents are due to obstruction of the outlets of sweat glands or keratomalacia and are often seen on the borders of the eyelid They give rise at times to irritation

which accompanies diphtheria membranous conjunctivitis however may be non diphtheritic Meningitis may cause inflammation and discharge perhaps unilateral similar to that caused by the gonococcus

Conjunctivitis may be a serious manifestation of tularemia (see Chapter 17)

The viral infections for instance acute coryza (head colds) influenza measles yellow fever and smallpox often have conjunctival manifestations The rickettsial diseases all affect the conjunctivae

Purulent conjunctivitis or ophthalmia neonatorum is usually caused by the gonococcus This is an extremely severe form of conjunctivitis and in the newborn child is a potent cause of blindness if not immediately treated by silver nitrate drops in the eyes

Gonorrheal conjunctivitis in the adult is acquired by direct transfer of material infected with gonococci to the eye The usual method of transfer is by the hands from the genitals The unusual transfers occur from the use of soiled towels or spattering of pus containing the organisms The incubation period is from a few hours to 2 or 3 days

The disease varies in severity In the virulent infections the eyelids may begin to swell and within a few hours become so tense that they cannot be opened In all there is a rapidly developing chemosis The lids and fornix of the affected eye are swollen tense and covered with a profuse purulent secretion In severe infections there may be only a blood tinged discharge In a few days the symptoms begin to subside but the eye continues to discharge pus for 2 to 3 weeks The course as well as the disease has changed greatly since the advent of chemotherapy and antibiotic therapy

Untreated or inadequately treated gonorrheal conjunctivitis often injures the cornea—corneal ulceration Ulceration is followed by cicatrization staphyloma and sometimes a panophthalmitis

The diagnosis of gonorrheal conjunctivitis is usually obvious from the history One eye alone is affected In the mild forms of the disease the diagnosis may be difficult because of resemblance of the disease to other forms of acute bacterial conjunctivitis and viral conjunctivitis particularly swimming pool or inclusion conjunctivitis The diagnosis is made from positive smears and cultures

The subjective symptoms of acute conjunctivitis are itching and smarting sensations usually referred to the eyelids These sensations become progressively more intense and the lids feel hot and heavy as though sand or foreign bodies were underneath the lids There is a varying amount of photophobia Owing to altered secretions on the cornea there may be blurring of the vision The symptoms are usually worse toward evening than during the rest of the day The affection may be limited to one eye but usually both eyes are implicated either from the start or within a very few days after the first eye is affected the other will become involved

All types of infectious conjunctivitis are characterized by the brilliant red color of the conjunctiva due to hyperemia and formation of new vessels When the upper lid alone is reddened the possible presence of a foreign body is a first consideration

The various types of *allergic conjunctivitis* are common and can often be distinguished by the character of the conjunctival secretions The secretion from allergic conjunctivitis particularly vernal catarrh has a high fibrin content and is stringy in character in contrast to the flaky or amorphous exudate of an ordinary bacterial infection

Allergic inflammations have a characteristically milky appearance This is often striking particularly in vernal catarrh and is useful in diagnosis It is probably due to the predominance of edema over cellular infiltration in allergic inflammations

A bulbar chemosis most frequently results from the instillation of too strong eye drops Full or excessive doses of many drugs such as arsenic potassium iodide and sulfonamides especially if there is an idiosyncrasy cause chemosis

Obstruction of the *nasolacrimal ducts* may be due to prenatal influence or may be the result of chronic nasal infection trauma or ulceration or may occur idiopathically Obstruction of the nasolacrimal ducts is manifested by epiphora at all times but is greatly increased by exposure to cold wind dust or smoke

Acute inflammation of the lacrimal gland is a rare condition The gland is sometimes enlarged in mumps or epidemic parotitis A bilateral enlargement of the lacrimal glands is rare *Dacryocystitis* infection of the lacrimal system may become chronic and produce a form of chronic conjunctivitis affecting chiefly the upper lid Occasionally a blepharitis accompanies these conditions Acute exacerbations of a chronic form of dacryocystitis may be manifested by acute swelling and obstruction resulting in abscess formation in the lacrimal sac

In *eversion of the puncta* the lower punctum looks forward and away from the depression where the tears accumulate and thus epiphora results Eversion of the punctum and the canaliculus may be of congenital origin or it may be caused by scarring from wounds or by chronic inflammation This condition most commonly occurs in old age or in facial palsy conjunctivitis blepharitis or ectropion The canaliculus may contain a cilium or concretions which irregularly obstruct the passages and produce a swelling of the tissue at the inner canthus of the eye

Malignant tumors of the lacrimal ducts and glands of the eye occur and these are usually sarcoma These tumors are classed with orbital tumors The most frequent orbital tumors are those that extend into the orbit from the adjacent structures usually from the meninges or the brain They may extend from the zygomatic and temporal fossae or from the nasal cavities When such infiltrations occur the eye is protruded and dry

Dermoid tumors arising from remnants of the orbital crest occur at the outer or inner angles of the eyes more frequently at the outer angles Dermoid tumors are to be differentiated from meningoceles

THE CONJUNCTIVA

The conjunctiva lines the eyelids (palpebral conjunctiva) and covers the eyeball (bulbar or ocular conjunctiva) in front

A *symblepharon* is a cicatricial attachment between the conjunctiva of the lid and the eyeball It is usually caused by an injury

A *pterygium* is a triangular fold of membrane occupying the interpalpebral fissure extending from the inner or outer part of the ocular conjunctiva to the cornea This condition occurs usually in elderly people who have lived in the wind and dust of the plains or deserts

Subconjunctival hemorrhage or ecchymosis often occurs without assignable cause Occurring after an injury either form of bleeding may indicate a fracture of the base of the skull anteriorly the blood passing forward into the orbit The other causes are the violent straining which may occur in severe coughing severe vomiting straining at the time of bowel movements and heavy lifting *Subconjunctival ecchymoses* may also occur during a convulsive seizure and in asthma or severe dyspnea They may be hemorrhagic infarcts and significant of septicemia or septicopyemia

Conjunctivitis or *inflammation* of the conjunctiva is common and often is caused by the presence of foreign bodies in the eye or by exposure to wind either hot or cold and to dust or pollens smoke or intense light

The infectious forms of conjunctivitis due to bacteria may be acquired from contact with others that have them usually these forms are limited to families Epidemics of conjunctivitis are caused by the pneumococci staphylococci and sometimes the *Hemophilus influenzae* An epidemic form of conjunctivitis probably of bacterial origin is often known as *pink eye* and is likely to occur during the spring or fall months (vernal conjunctivitis) Conjunctivitis may be of the membranous type

In some pseudomembranes are present on the conjunctiva of the lower lid On removal these pseudomembranes leave bleeding points

In about half of those who have epidemic keratoconjunctivitis the *second eye* becomes involved after a varying period of time As a rule the course of the disease in the second eye is much shorter and the inflammation considerably less than in the first eye

Systemic manifestations are headache and night pain fever and malaise which may occur with involvement of the one or the second eye Corneal involvement is a frequent and serious complication and this may occur without any signs of pain or photophobia or other subjective or objective symptoms The intensity and sequence of appearance of the symptoms therefore have no bearing on the duration or intensity of the disease Often eyes which show violent symptoms in the beginning may clear up rapidly and without resultant visual impairment The time interval between the onset of the disease and the involvement of the cornea varies from 1 to 5 weeks Usually the first sign of corneal involvement is the appearance of several tiny punctate infiltrates of the cornea in the subepithelial layer The corneal epithelium remains intact and does not stain with fluorescein In the majority of cases the corneal infiltrates are on the exposed part of the cornea that is within the confines of the palpebral fissure and usually in the pupillary area

The diagnosis of epidemic keratoconjunctivitis depends on the presence of an acute keratoconjunctivitis with practically no discharge from the conjunctiva This stage is followed by an acute swelling of the preauricular nodes and in some there is also swelling of the submaxillary and cervical nodes

Herpetic Keratoconjunctivitis The clinical characteristics which should arouse suspicion of an herpetic etiology in any inflammatory condition of the eye were outlined by Maumenee Hayes and Hartman as follows (1) the frequency with which it follows pyrexia (2) relapses or recurrences (3) slight if any purulent discharges (4) herpetic vesicles on the lids (5) bacteriologic sterility of the conjunctiva (6) rapid and complete healing as a rule (7) hypesthesia of the eye (8) a visible and slightly tender preauricular node Many of these clinical manifestations can be caused by the virus of epidemic keratoconjunctivitis

Inclusion Conjunctivitis (Paratrachoma or Swimming Pool Conjunctivitis) This disease is caused by a virus of large particulate size belonging to the psittacosis lymphogranuloma venereum group of organisms The conjunctivae are increased greatly in thickness and contain numerous basophilic cytoplasmic inclusion bodies which are morphologically identical with those of trachoma The disease occurs in infants and in adults

The incubation period is from 5 to 12 days In the infant the disease develops rapidly particularly in the lower lid and pseudomembranes may develop rapidly After 10 to 15 days the disease decreases in intensity but it lasts for months or for a year

In the adult the disease develops as a follicular conjunctivitis with scanty discharge and periauricular adenopathy As in the disease in infants the lower lid is more severely involved than the upper lid The disease in the adult is more chronic than that seen in the infant

The diagnosis of inclusion conjunctivitis in the infant is made most easily by remembering that this conjunctivitis is likely to commence 5 to 12 days after birth whereas gonorrheal conjunctivitis begins very soon after birth The lower fornix has a coxcomb like appearance and inclusion bodies can be demonstrated Trachoma never occurs in the newborn child

In the adult diagnostically significant is the presence of inclusion bodies from the lower lid in a patient who does not have pannus or significant conjunctival cicatrization The follicular hypertrophy is much more definite in the lower lid than in the upper

Ulcers of the cornea are attended with reddening of the eye Characteristically, glaucoma may cause reddening of the conjunctivae

Facial paralysis by causing undue exposure of the eyeball or a neuralgia of the fifth nerve may give rise to redness and inflammation

Follicles are not visible on the normal tarsal conjunctiva When follicles are visible they have hypertrophied The significance of *follicular hypertrophy* depends primarily on location and secondarily on their prominence Follicles involving the upper tarsal conjunctiva are suggestive of trachoma When the upper tarsal conjunctiva is clear and follicles in other parts of the conjunctivae are affected the diagnosis usually is follicular conjunctivitis The follicles of trachoma are soft gelatinous and easily expressed while the follicles of follicular conjunctivitis are hard and expressible only with difficulty

In a true *membranous conjunctivitis* coagulation of the fibrinous exudate has occurred so that removal of the membrane leaves a bleeding rough surface Conjunctival *pseudomembranes* are of two types the fine transient type and the thick relatively permanent type An example of a fine transparent membrane occurs in vernal catarrh acute conjunctivitis such as gonococcal conjunctivitis and epidemic keratoconjunctivitis

Thick opaque pseudomembranes occur in diphtheria in streptococcal infections in erythema multiforme and occasionally in ocular pemphigus

Conjunctival ulceration is characteristic of conjunctival tuberculosis and oculoglandular tularemia chancre of the conjunctiva and acute pemphigus The ulcers of acute pemphigus are readily distinguished from other ulcers by their shallow character and transient nature

The conjunctival *phlyctenule* or an elevated nodule or an ulcer of the corneal conjunctiva is pathognomonic of phlyctenular conjunctivitis and is believed to be an allergic reaction to a bacterial antigen commonly tuberculoprotein Unlike conjunctival phlyctenules corneal phlyctenules lack diagnostic value

Conjunctival granulomas are associated with buried foreign bodies or ruptured chalazia conjunctival tuberculosis and oculoglandular tularemia The conjunctivae in leptothrichosis have small whitish or grayish necrotic foci without gross ulceration A single large granuloma without diffuse conjunctival inflammation is suggestive of rhinosporidiosis

Conjunctival concretions indicate the occurrence of past or present chronic conjunctivitis Their diagnostic value is slight but when they occur in large numbers they can be considered suggestive of an old trachoma

Conjunctival *scars* follow injury operation for chalazia or burns trachoma pemphigus diphtheria and irradiation The scars of trachoma involve the upper tarsal conjunctiva predominantly and those of ocular pemphigus the lower tarsal conjunctiva and fornix Ocular pemphigus may cause an obliteration of the entire lower fornix a condition which distinguishes it from trachoma

The term *superficial punctate keratitis* is a descriptive name of epidemic keratoconjunctivitis and it is often employed as a general morphologic term for the designation of any condition presenting multiple small discrete lesions of the cornea affecting the epithelial and immediately adjacent subepithelial tissues

Epidemic Keratoconjunctivitis This is a disease of viral origin There is evidence to indicate a close immunologic relationship between the viruses of epidemic keratoconjunctivitis and St Louis Japanese and West Nile encephalitis viruses

The earliest symptoms vary greatly in intensity They commence with a sandy feeling redness and swelling of the palpebral conjunctiva of *one eye* At this stage it may be thought that there is a foreign body present causing tearing of the eye After 12 to 36 hours the bulbar conjunctiva too becomes congested and hyperemic and varying degrees of edema of the lids (particularly of the upper lid) ensue Occasionally the amount of edema is extreme and it may be difficult to open the eyes

be present along the lower retrotarsal fold without any evidence of inflammation and as such the condition is known as *folliculosis*

THE CORNEA

The cornea is the transparent portion of the external coat of the eyeball the anterior extension of the sclera The cornea consists of four layers (1) the corneal epithelium which is continuous with that of the conjunctiva (2) the substantia propria (3) the posterior elastic lamina and (4) the endothelium of the anterior chamber of the eye The last three layers anteriorly are continuous with similar tissues of the sclera However in the sclera there is not the definite arrangement of tissues as in the cornea In contrast to the cornea the outer surface of the sclera is bluish white and nontransparent

Diseases of the Cornea Many of the inflammatory diseases of the cornea have been discussed in connection with inflammation of the conjunctiva which is continued over the cornea Injuries of the cornea have been included with other injuries of the anterior part of the eye

Ulcers A reddened painful photophobic eye with a clear cornea except for a loss of substance at one point is an example of a corneal ulcer Corneal ulcerations often result from scratches or foreign bodies conjunctivitis and burns of the eyes Exophthalmos unilateral from a tumor of the orbit or as a progressive condition of unknown cause but usually associated with exophthalmic goiter may be the cause of corneal ulceration The ulcerations in exophthalmic goiter and also in some cases of facial paralysis may be due to the constant exposure of the eye consequent on the inability of the lids to cover the protruding eyeballs

The unilateral inflammation of the eye in meningitis may lead to a corneal ulcer In disease of the first division of the fifth nerve for instance herpes zoster with or without resulting anesthesia of the cornea and conjunctiva the cornea may undergo ulceration occasionally intractable

Hypopyon is a collection of pus in the anterior chamber of the eye which may accompany corneal ulceration The pus is derived not from the corneal ulcers but from an exudation from the iris and the ciliary body when these parts participate in the inflammation

Opacities of the Cornea Opacities of the cornea are usually caused by corneal ulceration or involvement of the cornea by inflammation or ulceration as the result of an injury An opacity of the cornea will reduce vision when it encroaches on the pupillary area Over the pupillary area a very slight opacity may cause much visual disturbance on account of the resulting diffusion and irregular refraction of light A linear or a nebular form of opacity will cause a great deal more visual disturbance than a circumscribed opacity Dense opacities or deformities of the cornea may cause a disfigurement of the eye as well as interfere with its vision

Interstitial or deep keratitis is characterized by a cellular infiltration of the deep layers of the cornea without ulceration during any stage of its existence There is involvement of the uvea which accounts for many of the symptoms

In many instances the disease is due to *congenital syphilis* It affects girls and boys alike and becomes manifest between the ages of 5 and 15 years In rare instances the patient may be older

During the early stages of congenital lues there are successive periods of photophobia lacrimation pain and interference with vision in both eyes In some instances the symptoms subside and the eye clears up entirely or nearly so

In the beginning there is present a patch of grayish infiltration which may be situated in any part of the cornea The patch enlarges to involve the entire front of the eye The cornea becomes softened The blood vessels begin to appear over the periphery of the dull gray appearing cornea giving the limbus a reddened color Vision may be reduced to light perception only However improvement will usually take place eventually

In both infants and adults the disease resolves without residual conjunctival or corneal changes

Trachoma Trachoma is caused by a filtrable virus The virus of trachoma is distinctive from most other viruses since it seems to be susceptible to chemotherapy to originate basophilic intrusion bodies and to have an intercellular cycle of morphologic variation For these and other reasons it has been suggested that trachoma and inclusion conjunctivitis should be classified with the rickettsial diseases An objection to such a classification is that trachoma is not transmitted by an arthropod host

Trachoma is a highly contagious disease which is responsible for much partial or total blindness The disease may present few or no symptoms The onset of the disease usually is with a mild conjunctivitis with thickening edema and congestion of the conjunctivae In some patients there are photophobia blepharospasm lacrimation itching burning and painful sensations accompanied by visual disturbances

On examination in the early stages of the disease there may be found minute follicles which can be seen in the upper tarsal conjunctiva On microscopic examination of the scrapings from these follicles the characteristic inclusion bodies may be found At this stage it is difficult to differentiate trachoma from inclusion conjunctivitis or gonorrheal conjunctivitis As the disease progresses there is papillary hypertrophy of the tarsal conjunctiva especially of the upper lid which imparts to the lid the characteristic granular appearance With further progression the papillae may increase in size and there appears a diffuse infiltration of the conjunctiva which is studded with grayish rounded translucent follicles arranged in parallel rows The trachomatous pannus consists of loops of blood vessels extending into the cornea between the epithelium and Bowman s membrane these loops are caused by a stopping or cutting off of the blood vessels by scarring The cornea presents a cloudy appearance and may be gray and translucent Its surface becomes vascularized the blood vessels stringing from the conjunctival vessels at the limbus This process advances until it covers the upper half of the cornea In severe cases it may cover the entire cornea and thus vision and perception of light are reduced and the patient loses his eyesight

From this stage of development of trachoma the disease may continue to progress or complete retrogression may take place so that the cornea again becomes transparent The final visual condition depends on the extent of corneal opacification and unevenness In severe instances of the disease iritis develops which is due to a change similar to those which occur in the conjunctivae Superficial corneal ulcers may appear near the margin of the advancing iritic pannus Ptosis may develop and become permanent

Trachoma after months or years reaches the stage at which formation of scar tissue is the predominating process As scar tissue forms the papillae and the follicles disappear In the tarsal conjunctiva there may have formed narrow bands of white scars which parallel the lid margin In general trachoma heals by the formation of scar tissue Consequently the sequelae are as follows ectropion entropion or trichiasis Often the movements of the eyeballs are restricted iritic pannus may be present and corneal opacities may result from the corneal ulcers Staphyloma of the cornea may follow a perforation of a corneal ulcer A dry scaly state of the conjunctiva may be a sequela of severe trachoma

The diagnosis is made from the prolonged course and on the basis of the clinical examination and the finding of characteristic minute inclusion bodies in the cytoplasm of epithelial cells in scrapings from the conjunctiva

Chronic Conjunctivitis Any of the acute forms of conjunctivitis may be followed by chronic conjunctivitis the symptoms of which are essentially a prolongation of an acute form of the disease *Follicular conjunctivitis* is a form characterized by the occurrence of lymph follicles distributed along the lower retrotarsal fold They appear as small round oval pinkish bodies arranged in rows These follicles may

mortar or whitewash strong acids and alkalies endanger the eye by injuring the cornea and producing symblepharon Strong ammonia is particularly harmful causing necrosis of the cornea hydrochloric acid much less so The central core of golf balls often contains caustics such as barium sulfate or sodium hydroxide which spurt into the eye if the ball is slit open

Ensuing immediately on the introduction of a caustic into the eye are intense conjunctivitis and chemosis but the cornea looks clear in this stage it is difficult to be certain of the extent of the injury A drop of fluorescein solution in the eye will reveal on magnification the extent of the area denuded of epithelium Therefore the patient is urged to secure the services of an ophthalmologist until the gravity of the injury can be determined In severe injuries the cornea is dull or opaque In the succeeding days an eschar forms and is thrown off This is followed by granulation of the injured conjunctiva and frequently by ulceration of the cornea In severe burns the foregoing process may cause the whole cornea to be destroyed perforation takes place and the eye shrinks In less severe burns a dense milky white opacity forms porcelain like in lime burns and sight is lost There is little danger of the loss of sight in the less severe injuries but the conjunctiva may form adhesions of the lid to the globe Adhesions are most likely to occur with the lower lid the caustic acting principally on the lower fornix which is obliterated by organization of the granulation tissue The symblepharon thus produced impedes the movements of the globe and may even interfere with its nutrition

THE ORBIT

Pain In and Around the Eye Foreign bodies and inflammatory diseases of the iris or conjunctiva are common causes of a painful eye Pain in and around the eye is frequently due to trigeminal neuralgia or to migraine Before treatment is given these conditions should be differentiated from acute glaucoma to avoid error in therapy The presence of dimmed vision reddened eye hazy cornea dilated pupil and greatly increased tension perhaps with nausea and vomiting will declare the case to be one of glaucoma

Photophobia is a form of eye pain excited by exposure to light It is present to varying degrees in the conjunctivitis of measles influenza the early stage of pertussis and severe coryza or the formation of the vesicles of variola and varicella on the cornea Ocular or retinal hyperesthesia is a rare form of photophobia not dependent on local inflammatory disease of the eye and it is usually although not always due to functional disturbance of the optic tract and nerve or the cortical centers for vision It is a frequent symptom in neurasthenia hysteria migraine and at times the hypnotic state It may be present in cinchonism in the early stages of the rickettsial fevers and in meningitis

Orbital Abscess Suppuration may originate within the orbit as from a general inflammation and suppuration of the eye which breaks the eyeball and spreads in the orbital tissues (panophthalmia)

Suppuration of the orbit may originate extrinsically and extend into it as from erysipelas from caries of the orbital bones or adjacent structures or in syphilis If pus enters the orbit it is usually from suppuration of the frontal sinus and ethmoidal cells If so the swelling presents at the upper portion of the inner angle of the eye Pus in the maxillary sinus usually discharges into the nose Pus within the orbit tends to distend the eyelids and to push the eyeball forward

Tumors of the Orbit Tumors may originate in the orbital contents as sarcomas of the lacrimal gland or the highly malignant choroidal tumors The majority of the orbital tumors however arise extrinsic to the orbit and enter through its natural openings or extend directly through its bony walls Tumors arriving in the orbit through natural openings may enter (1) from the brain through the optic foramen or sphenoidal fissure (2) from the region of the zygomatic and temporal fossae

In some instances all of the signs of the disease disappear In others when there is severe involvement of the iris choroid and vitreous sight may be lost or there may remain a permanent opacity in the cornea

The diagnosis of interstitial keratitis of syphilitic origin is based on the history The physical examination often reveals the presence of Hutchinson s teeth scars at the angles of the mouth where rhagades have healed and head deformities The head deformities consist of a square forehead and prominent frontal eminences Often there are enlarged lymph nodes in the posterior triangles of the neck There may be visible tibial deformities The roentgenographic examination of the bones may reveal irregular cortical outlines

The classic triad for the diagnosis of congenital syphilis is (1) interstitial keratitis (2) Hutchinson s teeth and (3) deafness (labyrinthine disease)

Arcus Senilis This is a degenerative condition of the cornea The true arcus is an ill defined grayish partial or complete ring at the circumference of the cornea the cornea itself being somewhat hazy It was formerly wrongly considered to be an indication of beginning arteriosclerosis and the degeneration of old age The false arcus is a sharply delineated ring of a clear yellow or yellowish white color due to a deposit of fat Neither one is of diagnostic significance

INJURIES TO THE ANTERIOR STRUCTURES OF THE EYE FROM FOREIGN BODIES AND PHYSICAL AGENTS

The eye is subject to injury from foreign bodies burns the action of caustics contusions wounds and radiant energy

Foreign Bodies Foreign bodies on the conjunctiva or on the cornea cause sudden discomfort and blepharospasm The foreign body may adhere to the palpebral conjunctiva or it is likely to be dragged across the cornea which it excoriates It may be floated by tears toward the inner canthus and pass into the nasal duct Very frequently it becomes lodged at about the middle of the upper sulcus subtarsalis where it is likely to irritate the cornea or in the upper fornix When it adheres to the bulbar conjunctiva it may lodge there Foreign bodies are generally imbedded in a mass of granulation tissue The chitin of insects and the husks of seeds may adhere to the cornea usually at the limbus for several days or even weeks or until the inflammatory reaction is sufficient to dislodge them

Particles of steel and emery are likely to penetrate the epithelium or substantia propria Larger particles of steel stone or glass may perforate the globe When situated in the cornea they cause great pain and irritation The pupil is often constricted If allowed to remain they expose the cornea to the dangers of infection by organisms in the conjunctival sac and ulceration This may lead to a small superficial slough being cast off carrying the foreign body with it The small ulcer thus formed may heal but if virulent organisms are present a spreading ulcer with or without hypopyon may develop

Foreign bodies are probably more frequent in the lower cul de sac than in the upper but in the former location they may be removed by the lacrimal fluid or by the patient s own efforts The upper cul de sac requires more scrutiny than does the lower, for it extends several millimeters above the tarsus in the upper lid and simple eversion of the upper lid may not expose it When a foreign body is suspected but remains undetected the tarsus should be everted and then the cul-de sac stretched forward and convexly with a lid everter The fold behind the lacrimal caruncle is another recess which may conceal a foreign body

Intra ocular foreign bodies are suspected in blast injuries or when the patient has been near the application of striking grinding or cutting force applied to metal

Burns and Injuries by Caustics Burns by hot water steam hot ashes exploding powder or molten metal and injuries by caustics such as lime usually from fresh

if toward the nose the strabismus is convergent or internal Vertical squint may be upward or downward By asking the patient to look at the tip of the examiner s finger it is usually easy to decide which eye squints and whether the squint is con vergent divergent upward or downward If the strabismus is slight and uncertainty arises the finger should be held directly in front of the face about 18 inches (46 cm) distant and the patient asked to gaze at it The apparently normal eye should then be covered by a card The uncovered eye if it be the squinting one will im mediately alter its position so as to fix itself on the finger, thus demonstrating that it had not before been properly directed

Strabismus with reference to its origin may be of two kinds concomitant and paralytic

In *concomitant strabismus* the squinting eye will retain its mobility moving readily in every direction concomitantly with its companion the degree of strabismus remaining substantially the same The principal causes of concomitant strabismus are overaction of certain muscles consequent on refractive errors and imperfect vision of an eye with resulting weakness of the muscles

In *paralytic strabismus* the squinting eye loses some of its power of movement owing to a loss of power in one or more of the external ocular muscles Closely con nected with and dependent on paralysis of the ocular muscles and strabismus is the subjective symptom of diplopia

Diplopia Double vision depends on the fact that unless the visual axes are correctly adjusted each to the other the images of the objects do not fall on identical points in the retinas Under normal conditions each image falls on the macula lutea of each retina combining to form a single image in the sensorium

Aside from the double vision which is produced by the action of alcohol atropine and other drugs on the ocular nerves diplopia like strabismus is a symptom of paralysis of the ocular muscles Any lesion of the brain spinal cord or basal meninges which causes paralysis of one or more of the external ocular muscles may be at tended with diplopia

Double sight with one eye may occur in cataract and astigmatism owing to irregular refraction and in brain tumor as a psychic aberration

Conjugate Deviation If both eyes are turned strongly and persistently toward one side either right or left the condition is termed conjugate deviation This con dition may be due to either spasm or paralysis of the internal rectus of one side and the external rectus of the other side and the eyes may be turned toward the lesion or away from it depending on its location and whether it is irritative or destructive This symptom depends on the normally associated action of the internal rectus of one side and the external rectus of the other in effecting horizontal co ordinated motion of both eyes to the right or left

The lesions producing conjugate deviation are in particular cerebral hemor rhage thrombosis tumors and meningitis Often if the lesion is a left sided one and causes right hemiplegia the eyes look toward the lesion but it is said that if the lesion is irritant or becomes so and spasms or convulsions occur the eyes turn away from the lesion

The determination of the nature of the lesion must be made by study of the associated symptoms of brain tumor meningitis apoplexy sclerosis or abscess

Nystagmus This is a rapid involuntary oscillation or slow movement of both eyeballs usually from side to side (lateral nystagmus) sometimes vertical or rotary It is a clonic bilateral spasm of the ocular muscles due to some irritation either functional or organic affecting the ocular muscle centers If nystagmus is very slight it may be intensified by directing the patient to look steadily at an object held well to one side or the other or if the nystagmus is vertical or rotary by directing the eye upward or downward or circumducting the eye by moving the object

Nystagmus is present in many cases of blindness or defective sight caused by

through the sphenomaxillary fissure or (3) from the nasal cavities through the nasolacrimal duct

Tumors which arrive in the orbit by penetrating its walls may come (1) from the nasal cavities and ethmoidal cells pushing through the thin internal wall, (2) from the frontal sinus appearing at the upper inner angle (3) from the sphenoidal cells at the posterior portion of the inner wall (4) from the brain cavity above breaking through the roof or (5) from the maxillary sinus below pushing through the floor

Dermoids and Meningoceles In the fetus the frontonasal process extends to join the maxillary processes on each side This leaves an orbitonasal cleft to form the orbit Owing to defects in the development of this cleft dermoid tumors may occur in its course They are seen at either the outer or the inner angle of the eye They are commoner at the outer angle near the external angular process than at the inner angle and may have a prolongation to the dura mater They also occur at the inner angle at the frontonasal suture At this point also meningoceles are likely to occur

Symptoms Indicative of Disturbances of the Ocular Muscles PTOSIS Drooping of the upper eyelid with inability to raise it (ptosis) may exist alone but it may be combined with paralysis of the third nerve Ptosis is usually due to paralysis of the levator palpebrae As the levator palpebrae is innervated by the second portion of the oculomotor nucleus ptosis is dependent on some interference with the function of the third nerve its nucleus or its cortical center

Ptosis may be congenital and hereditary unilateral or bilateral partial or complete In complete ptosis the lid drops enough to interfere with vision The attempts made to counteract this interference with vision consist of throwing the head back and raising the eyebrows by forced contraction of the frontalis muscle Instances of partial ptosis may be masked by these compensatory mechanisms Detection of partial ptosis is accomplished by asking the patient to look upward while the eyelashes are pressed firmly against the inferior bony margin of the orbit

Berke classified congenital ptosis in accordance with the muscles or structures involved separating them according to whether the superior rectus muscles are involved alone or there is weakness of the superior rectus and the inferior oblique muscles combined In addition to involvement of the recti and inferior oblique muscles attention is paid to ptosis associated with the jaw winking phenomenon and finally to ptosis that may be associated with blepharophimosis Congenital ptosis is most frequently unilateral This classification of congenital ptosis is useful since it is helpful in selecting the proper surgical procedure if any is to be employed

Acquired ptosis is usually unilateral It may be part of the symptom complex of paresis or paralysis of the whole of the third nerve or may be due to paresis or paralysis of the branch supplying the levator Isolated ptosis without other signs of oculomotor paralysis may result from disease of the brain (cerebral ptosis) Acquired ptosis may be due to direct injury of the muscle or its nerve supply deformity and increased weight of the lid brought about by edema trachoma and tumors In inflammatory necrotic processes and during marasmus with tissue loss the lid may sag from lack of support Bilateral ptosis may occur as a part of the syndrome of myasthenia gravis In severe neuralgia of the fifth nerve in tetanus in autonomic nerve lesions and in hysteria ptosis may coexist

The amount of ptosis sometimes alters with the position of the globe attaining its highest pitch in abduction of the eye its least in adduction In all forms of ptosis there may be a synkinesis of movement the lid rises when the jaw is moved as in mastication though it remains immobile when an attempt is made to look upward

STRABISMUS Squint or strabismus is inability to bring the visual axis of both eyes to bear at the same instant on one point the visual axis of one eye constantly deviating as a rule horizontally sometimes vertically from the object inspected If the squinting eye turns toward the temple the strabismus is divergent or external

lid from paralysis of the levator palpebrae and dilatation of the pupil and paralysis of the accommodations of the eye If the sixth or abducens nerve is paralyzed the eye cannot be turned outward If the fourth or patheticus nerve is paralyzed the superior oblique muscle fails to act and the double vision produced is worse when the patient looks down because the superior oblique is normally a depressor muscle The lacrimal frontal and nasal branches of the fifth nerve are nerves of sensation hence in supra orbital neuralgia and that affecting the nasal branch pain is felt in the orbit at the inner angle of the eye and down the side of the nose

Ocular Paralysis Paralysis of the ocular muscles is termed ophthalmoplegia In testing the ocular muscles for evidence of paralysis or weakness ask the patient to follow the finger moved in different directions Evidences of weakness or paralysis may thus be detected when there is inability of the eye to follow the moving finger If a more careful investigation is needed as with the slighter degrees of paralysis and the ocular asthenopias the patient should be referred to a competent oph thalmologist

Paralysis of the Third Nerve If there are ptosis slight exophthalmos external strabismus diplopia and a dilated pupil which reacts neither to light nor to accom modation and if when an object is moved in various directions the eye fails to follow it upward inward or far downward but will move outward and a little downward by means of the external rectus and superior oblique there is complete oculomotor paralysis In many cases some of the muscles escape the levator and rectus superior or the iris and the ciliary muscle being alone affected or ptosis may exist with a dilated pupil

The most frequent causes of oculomotor paralysis are exposure to cold and to syphilis the latter giving rise to a basal meningitis or a gummatous growth involving the roots of the nerve Various forms and degrees of oculomotor paralysis are significant symp toms in cases of encephalitis and botulism

Partial paralysis may be due to a cortical lesion affecting the inferior parietal lobule Undue exposure to light migraine and the excessive use of alcohol or morphine may cause temporary paralysis

As organic disease of the third nerve nucleus is usually associated with disease of the fourth and sixth nuclei an oculomotor paralysis without loss of power of the external rectus and superior oblique is as a rule functional

While the nerve is penetrating the crus it may be involved by tumor or other lesion of the crus in which case there will be partial or complete third nerve paralysis con joined with hemiplegia on the opposite side of the body a combination characteristic of unilateral disease of the crus If in addition there are unilateral paralysis and atrophy of the tongue the lesion is localized in the inner and inferior aspect of the crus involving the cerebral fibers passing to the nucleus of the hypoglossal or motor nerve of the tongue

Ptosis occurring on one side disappearing and then appearing on the other side (alternating) may be due to variable lesions in the neighborhood of the nucleus

At and after its emergence from the pons on its way to the sphenoidal fissure the third nerve may be involved by basilar meningitis or tumors

The lesions of locomotor ataxia which cause ptosis may be in the same locality If due to disease at the base the oculomotor paralysis is usually bilateral and likely to be accompanied by interference with the function of the other cranial nerves Third nerve paralysis may also be one of the symptoms of primary muscular atrophy and upper bulbar palsy

Oculomotor paralysis may be indicative of pressure in the cavernous sinus or inflam mation at the sphenoidal fissure or injury (fracture) in either locality

Loss of accommodation loss of light reflex and external squint following an attack of diphtheria are usually indicative of a neuritis of the third nerve

Paralysis of the Fourth Nerve The fourth nerve supplies the superior oblique muscle and paralysis of this muscle is somewhat difficult of detection If paralyzed when the fixing object is moved downward below the horizontal line the diseased eye fails to follow it there is slight convergent strabismus and the patient has double vision while looking down

optic atrophy amaurosis and corneal opacities It may be found in albinos It is frequently seen in epileptic or other convulsions and may form a part of the symptomatology of neurasthenia hysteria chorea and in some cases, of multiple sclerosis and insanity

The most important diagnostic associations of nystagmus however, are with Friedreich s ataxia disseminated (insular) multiple sclerosis and brain tumor especially growths involving the cerebellum pons or crus Acute basal meningitis is likely to present nystagmus

A special variety of nystagmus may be inherited It resembles in appearance the ocular type yet it is not associated with defective vision The movements of the eyes are constant horizontal and pendular of small range on forward gaze and of increasing amplitude on gaze to the sides The rate is about 120 per minute In most cases it is noted by the mother when the child is a few weeks to a few months of age

This type of nystagmus according to Rucker is usually a sex linked character the gene being carried on the X chromosome In some families hereditary nystagmus is transmitted on an autosome In some this type of nystagmus is associated with red green color blindness

IRREGULAR OR SPASMODIC MOVEMENTS OF THE EYES Nonrhythmic irregular movements of the eyes may occur in connection with special ocular paralyses or insufficiencies and errors of refraction Menière s disease when due to lesions of the labyrinth is sometimes accompanied by forced movements of the eyes Chronic hydrocephalus and meningitis (basal irritation) may give rise to irregular spasmodic movements of the eyeballs In hysteria the internal rectus and levator palpebrae may be tonically contracted the eyes being opened convergently squinted and uprolled

Asthenopia and Insufficiencies of the Ocular Muscles If complaint is made that close use of the eyes such as that involved in writing reading or sewing causes them to feel strained hot and uncomfortable and perhaps gives rise to headache in the forehead vertex or occiput the source of the trouble may be found in a weakness of the ocular muscles particularly the recti If this weakness exists the patient cannot keep the visual axes of the eyes in proper relation without an effort, either conscious or unconscious

If normal strength of the ocular muscles is present the condition is called *orthophoria* If some of the muscles are weak it is termed *heterophoria*

The varieties of heterophoria are *esophoria* weakness of the externi causing a tendency to convergence of the visual axes *exophoria* a tendency to divergence of the visual axes from weakness of the interni *hyperphoria* a tendency of the visual axis of an eye to deviate upward *hypophoria* the tendency of the visual axes to deviate downward

None of these deviations is sufficiently obvious to constitute strabismus All of these conditions require the advice of an ophthalmologist after he has examined the patient

The consensus among those trained in ophthalmology and neurology appears to be that the importance of muscular asthenopia in causing various forms of nervous disease has been greatly overrated According to the more conservative view muscular asthenopia is not a factor in causing nervous disorders but in neuropathic individuals it may intensify or render more frequent attacks of migraine trigeminal neuralgia occipital and cervical headaches vertigo and perhaps choreiform movements of the upper facial muscles The correction of refractive errors and the wearing of glasses may relieve the troublesome symptoms

Nerves of the Orbit The optic nerve is the nerve of sight Interference with it produces blindness This nerve will be discussed separately later in the text

The oculomotor or third nerve supplies all the muscles of the orbit except the external rectus and superior oblique If the third nerve is paralyzed the eye cannot be moved upward inward *or to any extent* downward There will be ptosis of the upper

The observations which may be made on examining the eyeball relate to pulsations in and around the eye the protrusion or recession of the eye its position and its degree of mobility

Visible pulsation of the eyeball may be found in aortic insufficiency arteriovenous aneurysm great cardiac hypertrophy and vasomotor instability as in exophthalmic goiter and other conditions in which there is abnormal throbbing of the arteries

Protrusion Exophthalmos of a moderate degree is commonly of congenital origin Protrusion of the eyeball when bilateral has as its commonest explanation exophthalmic goiter The eyes may protrude in asthma or other conditions attended by severe dyspnea Thrombosis of the superior longitudinal sinus may be attended by unilateral or bilateral protrusion of the eyeball and swelling of the face

Swellings or tumors may displace the eyeballs forward usually on one side perhaps on both sides as in great enlargement of the lacrimal gland and arteriovenous aneurysm exostosis cancer of the orbit or tumors of the upper maxilla One or both eyes may be pushed downward and forward and come to rest in the upper portion of the orbit by subperiosteal hemorrhages which may be present as one of the lesions of scurvy

Instruments to measure the protrusion of the eye have been devised the best known of which is the Hertel exophthalmometer a satisfactory instrument though somewhat difficult to use A simpler instrument is the one devised by Luedde but since the position of the two eyes is not measured at the same time it requires great care to be sure that the instrument is similarly placed on each side in turn The normal reading with the Hertel instrument seems to be about 15 mm but it is variable

The protrusion differs with different causes In a general overfilling of the orbit by increase in its contents the eye projects straight forward and can move in every direction This is the condition seen in exophthalmic goiter A tumor within the muscle cone if not too large produces the same effect but a large tumor in the cone tends to move and displaces the eyeball to the side Tumors in the orbit outside the muscle cone push the eye to one side

Malignant exophthalmos occurs in association with exophthalmic goiter The exophthalmos may increase despite relief from the toxic symptoms It has been postulated that this condition is due to a hormone different from the one which causes exophthalmic goiter

Various theories have been propounded to explain the development of exophthalmos by local reasons It has been variously attributed to an increase in the orbital fat to edema of the normal orbital contents to congestion of the vascular bed of the orbit to relaxation and hypertrophic myositis of the extra ocular muscles and to contraction of the smooth muscles Exophthalmos may follow paralysis of the ocular sympathetic nerves (see Autonomic Nervous System Chapter 23)

The malignant type of exophthalmos most commonly occurs in patients aged 40 years or more The sex incidence is almost reversed from that of the incidence of exophthalmic goiter Men are therefore more commonly affected than women Once a progressive or malignant exophthalmos is established rarely does a complete regression of the exophthalmos occur The condition may become stationary or it may progress until disorganization of the eyeball is so great that excision of the globe has to be performed It is now generally accepted that there is an edema of all the orbital tissues outside the globe and this increase in size of the orbital contents pushes the eyeball forward It also restricts the free movements of the eyeballs

Unilateral Exophthalmos Unilateral exophthalmos occurs as a nonpulsating and as a pulsating form

The most frequent causes of a *nonpulsating unilateral exophthalmos* are retrobulbar tumors of soft tissues bony orbital tumors or vascular lesions such as varicocele of the ophthalmic veins Infections of the sinuses soft tissues and bones

The diplopia is an extremely annoying symptom as the eye is so constantly employed in writing reading and walking all of these occupations requiring down turning of the eye The head is likely to be carried forward and toward the sound side

Isolated paralysis of the fourth nerve is very infrequent When present it may be indicative of pressure on the nerve trunk as it winds around the outer surface of the crus to reach the anterolateral edge of the pons

The coexistence of symptoms of cerebellar disease with fourth nerve palsy is indicative of a lesion of the anterior part of the cerebellar hemisphere on the same side There may be paralysis of the fourth nerve presumably due to a peripheral neuritis as the result of epidemic influenza diabetes alcoholism lead poisoning and tumor However such paralysis may be a transient symptom of myasthenia gravis

Sixth Nerve A lesion of the nucleus of the sixth nerve may cause conjugate deviation of the paralytic form

Third Fourth and Sixth Nerves Total ophthalmoplegia may be indicative of disease in the cavernous sinus of the sphenoidal fissure especially if there is anesthesia of the peripheral distribution of the ophthalmic division of the trigeminal nerve (skin of forehead upper eyelid and nose) The branches of this division accompany the ocular nerves through the sphenoidal foramen and they may be involved by lesions in these localities that is syphilitic or other periostitis gumma or other tumor aneurysm or arteriovenous aneurysm of the internal carotid or thrombosis of the cavernous sinus

Ophthalmoplegia external internal or total may be a symptom of acute or chronic polioencephalitis superior (upper palsy nuclear palsy) or encephalitis or botulism

When paralysis of all the ocular muscles is associated with facial paralysis without hemiplegia it is probably due to a basal lesion since the facial nerve like the ocular nerves is liable to involvement by disease at the base

DIAGNOSIS OF MULTIPLE OCULAR MUSCLE PARALYSES The diagnosis of paralysis of a single ocular muscle may be difficult on occasion When more than one muscle in the same eye is paralytic a much more difficult task is encountered and if one or more muscles in each eye are paralyzed the signs can be confusing owing to all degrees of paresis of the muscles and innervational overaction and underaction spasm contractures and fibrosis of other muscles not involved It can be safely said that only experienced ophthalmologists are aware of the difficulties that sometimes attend the analysis of ocular muscle paralysis

The ophthalmologist employs the Maddox rods screen test test of rotations or ductions test of versions test of diplopia fields past pointing and head tilting in his diagnostic procedures

Immediately after paralysis is acquired all the characteristic symptoms and signs such as diplopia limitation of motion primary and secondary deviation false orientation and the subjective sensations of vertigo and dizziness may be present As the paralytic condition continues the original symptoms become modified diplopia becomes less obtrusive compensatory head tilting becomes apparent fixation may change and unopposed or innervationally overacting muscles may become spastic and undergo contraction with fibrosis The originally nonconcomitant strabismus gradually takes on the characteristics of a concomitant disturbance The complexity of this transition is multiplied by the simultaneous paralysis of two or more muscles

POSITIONS OF THE EYEBALL

The positions of the eyeball in its socket are classed as flat midprominent or protruding With age the increasing relaxation of the extra ocular muscles causes the eyes to appear flat or less deeply imbedded than in early life This change is less marked in women than in men

of the wound become yellow and necrotic hypopyon appears there is chemosis with intense ciliary and conjunctival congestion and the lids are swollen and red The vitreous becomes purulent as shown by a yellow reflex by oblique illumination The anterior chamber soon becomes full of pus and the cornea cloudy and yellow ring infiltration may occur There may be exophthalmos and limitation of movement of the globe due to extension of the inflammation In the early stages of metastatic panophthalmia rapid failure of vision a yellow reflex and hypopyon are found

If the disease is permitted to take its course the sclera is ruptured usually just behind the limbus The pain subsides and after prolonged suppuration the eyeball shrinks and vision is lost Sympathetic ophthalmia is unlikely

As soon as it is evident that the eye cannot be saved it should be excised

Sympathetic Ophthalmia Sympathetic ophthalmia is a rare condition in which serious inflammation attacks the sound eye after injury of the other eye The disease is thought to arise from an infection

Sympathetic ophthalmia almost always is caused by a foreign body which remains within the eye without suppuration Wounds involving the iris ciliary body or lens capsule without suppuration are likely to set up sympathetic ophthalmia Perforating ulcers very rarely cause it

Children are particularly susceptible but the disease occurs at any age It usually begins 4 to 8 weeks after the injury to the first eye (the exciting eye) has taken place rarely earlier but the onset may be delayed for many months or even years

The manifestations in the second or sympathizing eye are almost always a plastic iridocyclitis in rare instances a neuroretinitis or choroiditis In other cases the patient first seeks advice for defective vision or inflammation in the uninjured eye (sympathetic irritation)

The symptoms commence with sensitiveness to light and transient indistinctness of objects Objects become blurred if the eye is fatigued but after an interval of rest vision improves When fully developed all the symptoms of iridocyclitis are present varying in degree according to the severity of the disease The prognosis as to vision is always doubtful but if there is much deposition of plastic exudates in the pupillary area it becomes extremely grave

On examination at this stage there may be lacrimation slight ciliary injection tenderness of the eyeball as shown by the patient shrinking from an attempt at examination precipitates on the back of the cornea and vitreous opacities These early symptoms may occur in intermittent attacks and spread over a considerable period As the disease progresses the signs of iridocyclitis are present

The treatment of sympathetic ophthalmia is one of the most difficult problems in ophthalmology and often demands the exercise of superior judgment on the part of the ophthalmologist When restoration of sight is doubtful the eyeball may have to be sacrificed if treatment fails

INJURIES OF THE EYEBALL

Wounds of the conjunctiva as has been said are common They usually heal readily Polypoid masses of granulation tissue sometimes form on the surface during the healing process A wound of the lens may escape notice at first especially if the wound is small Later the wound is made evident by the ensuing corneal opacity

Small wounds of the cornea limited to the center heal well unless they become infected When infections supervene a permanent dense opacity is left and the contraction of the organizing scar tissue may cause an irregular astigmatism

The danger of corneal opacities is greatly increased by a large wound of the eye if it extends into the sclera or if the lens is also wounded In the larger wounds of the sclera prolapse of the iris is almost certain to occur A physician should never yield to the temptation to replace a prolapsed iris even if this is possible since it may carry infection into the eye It must be excised

of the orbit may produce an exophthalmos Prominence of the eye may occur as a result of an anterior staphyloma or a high myopia

In the majority of instances *pulsating exophthalmos* is unilateral The condition usually follows trauma and is the result of a fistula between the internal carotid artery and the cavernous sinus In some instances a fracture through the base of the skull involving the body of the sphenoid may be demonstrated Spontaneous instances may occur from rupture of the artery within the sinus caused by arteriosclerotic plaques miliary septic aneurysms or congenital weakness of the vessel wall

The internal carotid artery lies within and is entirely surrounded by a venous channel This arrangement may permit the formation of arteriovenous fistulas with a minimum of trauma The pulsating exophthalmos resulting from an arteriovenous fistula in the cavernous sinus is due to a sudden engorgement of the venous space with arterial blood under high pressure There is a retrograde flow into the ophthalmic veins with transmitted arterial pulsations and a venous engorgement of the orbital contents and eyelids These changes may occur to a less degree on the opposite side as a result of communications between the two cavernous sinuses

The diagnosis of unilateral exophthalmos is usually easy depending on the degree of proptosis Pulsation does not always exist in those cases which occur spontaneously Following head injuries a bruit thrill and pulsation usually are present There are headache disturbance in vision and intracranial noises The sounds may vary from minor sensations to an almost continuous roaring The visual field may show a wide range of disturbance Diplopia may result from an increase in internal or extra ocular pressures The presence of chemosis accounts for some of these changes Blindness may be due to atrophy of the optic nerve to glaucoma or to cataract

The extrinsic movements of the eye are limited by the proptosis and the alterations in the nerve mechanism of the extra ocular muscles The retinal findings are not constant and depend on the duration of symptoms Compression of the common carotid artery will result in a cessation of a bruit thrill and pulsation when present Arteriograms may be obtained by the injection of thorium dioxide into the carotid arteries This procedure is useful in diagnosis in obscure cases The radioactivity of this or similar substances should limit its use

Recession Enophthalmos or sinking of the eyeball into the orbit if bilateral may be due to absorption of the cushion fat of the orbital cavity such as takes place in prolonged and continued fevers and in cachexias from any cause It is notably present during severe dehydration and shock It may also be present in consequence of a rapid and exhausting drain on the body fluids as in cholera severe diarrhea and large hemorrhages In collapse it may be due to a deficient orbital blood supply

INFECTIONS OF THE EYEBALL

Panophthalmia Exogenous and endogenous forms of panophthalmia are recognized *Exogenous panophthalmia* is generally caused by infected wounds whether accidental or the result of operations and by ulcers The vitreous is usually first affected organisms grow in it and purulent cyclitis retinitis and choroiditis result In most instances the deeper parts of the vitreous become infected

Endogenous panophthalmia arises during the course of septicemias or pyemias The small or the larger arteries in the eyeball may become occluded by septic emboli

The pneumococcus often is responsible for panophthalmia but the disease may be caused by staphylococci streptococci *Escherichia coli Pseudomonas aeruginosa Clostridium perfringens* and by saprophytic organisms such as *Bacillus subtilis*

In both exogenous and endogenous forms there is fever (temperature 102 to 103 F) headache drowsiness and sometimes vomiting There is severe pain in the eye due at first to iritis later to increased tension In the exogenous forms the edges

has been done to vision and unless the process of removal will almost inevitably destroy sight Magnetizable foreign bodies are more easily removed than others if the small or the large electromagnet is available

If the reaction in the eye does not subside in the course of a week or 10 days as shown by diminution of ciliary injection and cessation of photophobia and lacrimation the ophthalmologists may advise removal of the eye In the interval the cornea is examined each day by oblique illumination and magnification in search for precipitates The presence of these is a more definite indication for excision of the eye

In the presence of considerable prolapse of vitreous as well as of iris and ciliary body or if the lens is wounded there is little probability of saving the eye If it is almost certain that useful vision will be lost the risks of sympathetic ophthalmia should not be run and the ophthalmologists will advise prompt excision of the eye

GLAUCOMA

Glaucoma is a disease accompanied by increased intra ocular tension which results in hardness of the eye atrophy of the retina cupping of the optic disk and blindness It is supposed to be due to interference with the drainage of the eye and the circulation of the aqueous humor Glaucoma may be acquired may occur in families or may occur sporadically Glaucoma of acquired origin as a rule is a disease of advanced life The familial glaucoma occurs usually before the age of 40 years Instances of sporadic glaucoma too may affect middle aged persons The cause of the disease often is not known Perhaps the most definite thing recognized in its etiology is that there often is a hereditary tendency The disease tends to occur in hyperopic eyes myopic eyes are far less liable to the disease but they are not entirely exempt There are a number of varieties of glaucoma distinguished by varying symptoms

Acute glaucoma usually is preceded by a number of transitory attacks during which there is diminution in the acuteness of vision The sight appears to be obscured as by a fog The patient sees a ring or a rainbow of tints around lights

The cornea especially in its center will often show some congestion that is it has a clouded appearance There is a feeling of dullness or slight pain in the eyes and head The pupil is dilated and sluggish in action and the tension of the globe is slightly to moderately increased

The *acute glaucomatous attack* commences suddenly and violently in one eye during the early hours of the morning The constitutional disturbance may be so great that the patient is prostrated with an irregular intermittent pulse pallid face and cold extremities

The pain radiates throughout the entire head and is most intense in the trigeminal nerve Its acute onset even though associated with nausea differentiates glaucoma from intracranial conditions The pain is due primarily to pressure on the ciliary nerve and ciliary plexus

Edema of the eyelids is often present Lacrimation is an important finding especially in the absence of any external cause and is primarily due to reflex irritation In acute inflammatory glaucoma as contrasted with chronic compensated glaucoma the increase of intra ocular pressure is readily determined by palpation

In severe attacks of glaucoma the entire bulbar conjunctiva and sclera present engorged and dilated vessels so uniform that the color is a deep red This congestion is primarily a venous stasis from the increased intra ocular pressure In some cases minute hemorrhages may be seen

The cornea presents a steamy appearance Later the epithelium becomes elevated and is edematous The underlying stroma is edematous and is more or less translucent Edema is followed by vesicle formation involving the superficial epithelium

In an attack of inflammatory glaucoma the anterior chamber is shallow After the iris comes in contact with the posterior surface of the cornea adhesions rapidly

Occasionally in perforating wounds pyogenic organisms are carried into the eye and cause rapid dissolution of the cornea and panophthalmia As the result of the panophthalmia the whole of the central part of the cornea is cast off The infecting organism has generally been found to be *Pseudomonas aeruginosa* (*Bacillus pyocyaneus*)

Wounds of the corneosclera are particularly likely to originate sympathetic ophthalmia

When perforation of the sclera has occurred there is reduction in the intra ocular tension If the perforation is near the cornea the anterior chamber is shallow or obliterated If the wound is large prolapse of some of the contents of the globe occurs The uvea is easily recognized on account of its pigmentation since the uvea is the iris ciliary body and choroid considered together Very often the gelatinous vitreous can be seen hanging out of the wound Blood in the anterior chamber and vitreous hemorrhage may be present with or without perforation

An eye containing a foreign body is likely to be affected by sympathetic ophthalmia When there is present a perforating wound of the eye it may be difficult to be sure that it does not contain a foreign body The foreign bodies most likely to penetrate the eye and be retained are slivers of steel lead stone and particles of glass Parts or all of copper percussion caps and less frequently spicules of wood may penetrate the eye

A foreign body large enough to be seen by the unaided eye will cause so much damage that the eye may have to be removed Very small particles which penetrate the cornea or sclera and lodge in the deeper parts of the eye are revealed by roentgenographic study Most intra ocular foreign bodies are composed of metallic fragments and are thus radiopaque With the exception of glass containing lead in its manufacture glass does not cast a shadow on the roentgenogram

Depending on the nature of the foreign body the pathologic condition set up in the eye is profound and varied For instance copper causes suppuration of tissues and a leukocytosis even in the absence of pyogenic organisms This is due to the peculiar chemical irritation of copper Copper in the lens however may cause little reaction because the lens is destitute of active blood circulation Metals other than iron and copper—for instance lead zinc gold and silver—appear to cause little chemical reaction and usually remain quiescent

The foreign body may pass through the cornea or the sclera and originate minimal symptoms If it has passed through the cornea the wound or scar can be found by examination with oblique illumination and magnification

A very small foreign body may be retained in the lower part of the anterior chamber and be hidden by the hemispheric shape of the sclera Glass is particularly difficult to locate in the anterior chamber because its refractive index differs so little from that of the surrounding medium

When a foreign body passes into or through the lens a traumatic cataract is produced which undergoes the usual changes A hole in the iris is of great diagnostic significance for it indicates usually a perforation by a foreign body The foreign body may be visible in the lens either before or after dilatation of the pupil It is possible for a foreign body to pass through the iris and through the circumlental space without wounding the lens

A foreign body retained in the vitreous or retina when the lens is clear and there has been little hemorrhage may be seen ophthalmoscopically Often the foreign body can be located by the track it leaves through the vitreous The track is indicated by a gray line The foreign body generally black and often with a metallic luster, is surrounded by white exudate and red blood clot If the particle has been long in the eye it may have become encapsulated Particles more than 1 to 2 mm in size are almost certain to lead to the destruction of the eye

All foreign bodies should be removed by an ophthalmologist unless little damage

perforation may occur relatively early along the perivascular spaces of the ciliary vessels The orbital tissues then become infiltrated The lymph nodes are not affected metastasis occurs mainly to the liver but may be present elsewhere The first manifestations of metastatic sarcoma of the eye may appear months or years after the eye has been removed and may be revealed by enlargement of the liver and by jaundice

Sarcoma of the choroid appears in persons between the ages of 40 and 60 years It is always primary and involves one eye only

The symptoms commence with a defect in the field and diminution in sight depending on the exact seat of the tumor As the tumor enlarges there is pain in the eye The symptoms are produced more quickly when the tumor is near the macula than when it is peripheral since vision is affected early In other cases the tumor has usually attained a considerable size and the patients may seek treatment for relief of the pain of glaucoma

The cause of the glaucoma is obscure in some cases it is due to the lens and iris being forced forward so that the angle of the anterior chamber becomes blocked In other instances particularly those of rapid onset obstruction to the venous outflow from the eye is the probable explanation the tumor being in some instances so situated as to press on the venous outlet

On examination sarcoma of the choroid may resemble in its manifestation a primary detachment of the retina or glaucoma From primary glaucoma sarcoma of the choroid is distinguished by the fact that sight is involved before the inflammatory symptoms appear there are no prodromal symptoms such as usually precede glaucoma nor remissions in symptoms one eye only is involved the characteristic field of vision in glaucoma is not present and the unaffected eye presents no symptoms of glaucoma If the detachment of the retina is extensive differential diagnosis may be difficult under such circumstances the eye will be blind

Secondary carcinoma of the choroid occurs sometimes in late stages of scirrhous carcinoma of the breast more rarely in cancer of other organs

THE COATS OF THE EYEBALL

The outer strong fibrous coat of the eyeball is the sclera (sclerotic) The vascular middle coat is the choroid The retina or inner coat is the end organ of sight and is but an extension of the optic nerve (Fig 4 5)

The sclera forms a firm protective covering or case for the delicate retina within It is continued forward over the front of the eye as the cornea It is essentially a strong membrane which provides protection The common diseases of sclera and cornea are those of weakness If the cornea weakens it bulges forward and the protrusion is called an *anterior staphyloma* If the posterior part bulges toward the orbit a *posterior staphyloma* is formed

The cornea has no blood vessels but from its exposed position it becomes inflamed (keratitis) and ulcerated and eventually blood vessels may develop into it from its periphery constituting the vascularization known as pannus The affections of the cornea have been considered earlier

The weakest portion of this covering for the eyeball is at the junction of the sclera with the cornea On this account blows on the eye cause it to rupture usually at this point the tear encircling the edge of the cornea for a variable distance (usually at its upper and inner quadrant) according to the force and direction of the injury On healing a staphyloma may form at this point

Yellow sclera is observed in jaundice and carotenemia or it may be due to fatty deposits Bluish white or pearly sclera is found in the anemias in phthisis in nephritis and by contrast with the discolored face in Addison s disease There is a rare familial type of disease in which successive generations contain members who have fragile bones among other members who are healthy the latter have white scleras Those who have fragile bones have scleras which are definitely blue in color

form within 24 hours After 36 to 48 hours these adhesions become so dense that it is often impossible to break them except by operation The dilated often irregular and rigid pupil is one of the most characteristic objective symptoms in acute congestive glaucoma

In a severe attack of acute inflammatory glaucoma the vision is severely reduced often to light perception No part of the fundus can be seen with the ophthalmoscope If the condition progresses the increased tension of the eyeball causes an extravasation into the optic nerve disk which results in blindness

Often the ophthalmologist discovers a simple glaucoma the diagnosis of which is made by observing the acuity of vision charting the visual field making repeated estimations of the tension and interpreting the findings presented when the ophthalmoscope and the slit lamp have been employed in the examination Simple glaucoma is often difficult to discover because tension of the eyeball is not always present at the time of the examination the tonometer may reveal a rise in tension only after repeated tests on different days Progression of the disease destroys vision

The patient complains of dimness of vision with the appearance of concentric rings of color surrounding the source of bright light for instance a street light

TUMORS OF THE EYEBALL

The irides may contain dark brown spots (melanomas) due to congenital aggregations of retinal pigment epithelium As a rule these are benign but occasionally they take on malignant proliferation Any increase of size of such spots in middle aged persons causes anxiety to the ophthalmologist

True intra ocular tumors are rare When they are present they are of two varieties namely retinoblastomas or gliomas of the retina and sarcomas of the choroid The sarcomas of the choroid are frequently referred to as malignant melanomas

Retinoblastoma The retinoblastoma commonly known as glioma of the retina probably always congenital occurs in children less than 5 years old usually in one eye but often in both occasionally in successive children of the same family The attention of the parents is attracted by the yellow reflex easily seen through the pupil which is usually dilated

The ophthalmoscope shows small whitish or yellowish plaques (calcium deposits) with metallic luster either covered by detached retina or growing into the vitreous the surface often presenting newly formed blood vessels and sometimes hemorrhages

In the glaucomatous stage there are pain increase of tension and other symptoms of glaucoma The tumor increases in size and extends into the vitreous Very soon the growth can no longer be seen on account of turbidity of the media

As the tumor increases in size there are enlargement of the globe and exophthalmos and then perforation takes place The growth passes backward along the optic nerve to the brain and forward through the cornea and sclera increasing in size rapidly involving all tissues with which it comes in contact and forming a large vascular and ulcerating mass Metastasis occurs to the neighboring lymph nodes and to the cranial bones and becomes widespread causing death

Sarcoma Sarcoma of the eye arises in the iris choroid and retina Sarcoma of the *iris* grows rapidly extends to the corneosclera perforates the globe and gives rise to metastasis Sarcoma of the *choroid* arises from malignant proliferation of the stroma cells of the outer layers It forms at first a mass raising the retina over it By the process of growth increased tension is produced and rupture occurs in the subretinal space The *retina* remains in contact with the tumor at the summit of the head but is detached from the choroid at the sides As the tumor progresses the retina is more and more detached until no part remains in situ The lens then becomes opaque The tumor may fill the globe before perforating the sclera or

Diseases of the Iris and Ciliary Body The iris the ciliary body and the choroid form the second coat of the eyeball The iris is in front the ciliary body is contained in the folds of the choroid which forms with the lens the partition between the anterior and posterior chambers of the eye All these structures contain blood vessels and thus are the source of the nourishment for the eye These structures together comprise the uvea or uveal tract Disease affecting one of these structures is likely to involve all of them and may be termed uveitis

The *iris* is a circular membrane with a circular opening the pupil which serves to regulate the amount of light passing through the lens to the interior of the eye The iris lies on the anterior capsule of the lens when the pupil is contracted and it hangs free of the lens during maximal dilatation In treatment of disease conditions of the uvea advantage is taken of these responses of the iris

The color of the iris depends on pigmented cells of its stroma and on the cells of the retinal pigmented layers The muscles of the iris are the sphincter pupillae (third cranial nerve) and the dilator pupillae (sympathetic innervation)

The *ciliary body* is contained within that portion of the tunica vasculosa stretching from the anterior part of the iris to join the choroid It is composed of the ciliary muscle and the ciliary processes blood vessels and nerves

The blood supply is derived from the greater circle of the iris and the anterior ciliary arteries Some of the veins pass posteriorly to join those of the choroid while others from the ciliary muscles pierce the sclera and course beneath the conjunctiva

The ciliary processes are meridianally arranged vascular folds forming a circle These vascular folds supply nutrient fluids to the interior of the eye especially the cornea and lens The ciliary muscle is the muscle of accommodation When it contracts it draws the ciliary processes and choroid forward and inward thus allowing the suspensory ligament to relax and the lens to become more convex The ciliary body contains within its substance a nerve plexus with ganglion cells

The congenital malformations of the iris include pupillary membranous changes Normally the membranous covering of the pupil is absorbed but part may persist Of the congenital malformations of the iris the common one is *coloboma* a notching in the inferior half of the iris and often associated with coloboma of the choroid

Occasionally the two irides will be of different colors or a part of an iris may have a different color from the rest This condition is known as *heterochromia* It may be due to iritis but more frequently it is congenital

Loss of Power of Accommodation In paralysis of the ciliary muscle distant objects can be clearly seen but near objects are indistinct This condition must not be confused with (1) reflex iridoplegia (loss of the pupillary light reflex) or (2) accommodative iridoplegia (loss of the pupillary contraction for near vision) Reflex iridoplegia and accommodative iridoplegia both depend on the integrity of the same portion of the oculomotor nucleus Cycloplegia is one of the symptoms of complete third nerve paralysis is a common symptom in diphtheritic paralysis and may be present in multiple sclerosis

Iritis Iridocyclitis and Uveitis In consequence of its vascularity the iris is a frequent site of inflammation When inflamed it may adhere to the lens behind forming a posterior synechia An anterior synechia is an adhesion in which on account of a perforation of the cornea the iris washes forward and becomes attached to the cornea in front

Inflammation of the iris is so frequently associated with inflammation of the ciliary body that in most cases these two conditions can be distinguished *only by an* experienced ophthalmologist Consequently here this condition will be described as *iridocyclitis* which means an inflammation that involves the iris the ciliary body and often the choroid

The causes of iritis and often iridocyclitis if the involvement is more pronounced and accompanied by a more intense pericorneal injection tenderness *in the ciliary* region swelling of the upper lids turbidity of the aqueous and increased tension

Diseases of the Sclera Diseases of the sclera are not common The most frequent disease of the sclera is inflammation which may be either superficial or deep The superficial inflammations of the sclera are generally of the lymphatic part Scleritis however an inflammation of the sclera involving the entire thickness is a serious condition characterized by severe pain frequently extending to the neighboring parts of the eye and to the forehead In this condition the tension of the eyeball often becomes increased and a secondary glaucoma ensues On examination there are discernible well marked violet patches or dark red patches *adjacent to the cornea* If uveitis as is often the case accompanies the scleritis vision is often interfered with and may sometimes be lost entirely Thinning and bulging of the sclera are known as staphyloma of the sclera

Diseases of the Choroid The choroid or vascular coat of the eyeball extends forward and is folded at the separation of the anterior chamber of the eye from the posterior chamber

Coloboma Coloboma of the choroid is a congenital defect of the choroid and retina which is manifested by a large white patch bordered with pigment usually situated below the disk The white patch is the exposed sclera Retinal vessels pass across this area There is a scotoma (or scotomas if the condition is bilateral) corresponding to the defect or defects

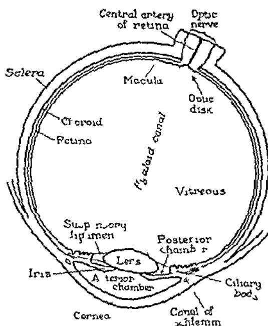


Fig 4 5 Coats of the eyeball and associated structures

Inflammation Inflammation of the choroid or choroiditis may be suppurative or exudative Suppurative choroiditis occurs when the choroid with other ocular structures is involved in a purulent inflammation usually extending from the orbit

The common form of choroiditis described by the ophthalmologist is the exudative form and properly termed chorioretinitis It may be caused by syphilis or tuberculosis but usually no cause can be assigned This condition does not present any external symptoms or signs The patient complains of disturbances of sight diminution in vision due to opacities in the vitreous distortion of objects and a reduction or loss of vision in that part of the field which corresponds to the area of exudation Very often the patient complains of seeing a black spot which is the manifestation of a positive scotoma There are often flashes of light sparks or bright circles before the eyes

If an area of the retina which is concerned with peripheral vision is involved the defect of vision may not be noticed but if the macula is involved the complaint will be a marked reduction or loss of central vision

On ophthalmoscopic examination the vitreous often is hazy and patches of exudation varying in size shape and position are reported by the ophthalmologist The areas of exudation are colorless with ill defined margins There is elevation of the retinal vessels which pass over these areas In mild instances the exudation may be come absorbed More commonly however the exudate becomes organized and owing to thinning of the areas of exudation the whitish sclera shows through Through these thinned atrophic areas choroid vessels surrounded by black pigment especially at the margins are reported

responding half of the optic nerve When the nerve reaches the optic chiasm it splits into two parts one (internal fibers) going to the opposite side of the brain and the other (external fibers) to the ganglia on the same side of the brain Posterior to the chiasm the nerve fibers form the optic tracts The optic tracts after leaving the chiasm wind around the crura cerebri to the external geniculate bodies thence they pass to the thalami and anterior corpora quadrigemina and are continued backward into the cuneus lobule of the occipital lobe of the brain

The retina is an extension of the optic nerve dispersed over the whole inner part of the lining coat of the eyeball Where the optic nerve begins this dispersion is termed the disk In the visual fields there is a blind spot corresponding to the disk Away from the disk are the macula lutea and fovea centralis or region of most distinct vision

Hereditary Degeneration In pigmentary degeneration of the retina morbid heredity is definite The characteristic may be dominant recessive or sex linked recessive The disease occurs from soon after birth to and through middle life

The ophthalmologist describes first a narrowing of the field of vision and twilight and night blindness The optic nerve head is usually pale There are deposits of dark pigment irregular in size and shape connected by fine pigmented lines The pigmentation is midway between the extreme periphery of the retina and the region of the macula and is bilaterally symmetric *The macular region is the last part of* the retina to be involved The disturbances of vision correspond to the retinal lesions The retinal vessels are reduced in size The retina is atrophic and choroidal vessels are exposed The progress of the disease is slow and there are no remissions

Congenital deaf mutism polydactylism defect of genital development mental defect convulsions epilepsy and obesity like that seen in Frolich s syndrome are often associated with pigmentary degeneration (see Chapter 15)

The diagnosis is usually made with ease by an experienced ophthalmologist

Acquired Degeneration The retina may undergo a chronic progressive degeneration consisting of atrophy of the retina with characteristic deposits of pigment Along with or as a part of general retinitis there may be a *macular degeneration* The condition is bilateral the maculae are the seat of pigmentary changes and central vision is reduced This commonly occurs in elderly people

Obstruction of the central vessels of the retinas causes immediate blindness If the artery is obstructed the retina is pale with a cherry red spot at the fovea If the vein is obstructed there are congestion and enlarged tortuous veins present everywhere in the fundus

Retinitis Inflammation of the retina is rarely limited to the retina alone but usually is associated with disease of the choroid and is thus termed chorioretinitis Inflammation of the retina may be associated with inflammation of an optic nerve and if so is known as neuroretinitis

As retinal disease is usually secondary to some extraocular cause it is in the majority of cases a bilateral affection The varieties and diagnostic significance will be indicated in various parts of this text A partial listing is as follows

Neuroretinitis of hypertension occurs in cases of arterial hypertension (see Hypertension Chapter 10) The papillitis may be so marked that it is difficult to determine whether it is of vascular or of intracranial origin In neuroretinitis the retinal vessels are altered and seem interrupted where they are covered by the swelling The arteries are thin the veins are tortuous and distended There are flame shaped hemorrhages and the so called cotton wool exudates *Neuroretinitis of glomerulonephritis* is distinguishable from that occurring in hypertension (see Diseases of the Kidneys Chapter 11) In *congenital syphilis* retinitis pigmentosa may occur In acquired syphilis both retina and choroid are likely to be affected and the chorioretinitis is regarded as a secondary symptom About 1 of every 4 who have leukemia has retinitis (*leukemia neuroretinitis*) *Diabetic retinitis* seems to be characteristic (see Diabetes Mellitus Chapter 14)

The symptoms of affections of the retinas are marked disturbances of vision or

are the same The distinction in the terms is based rather more on degree of involvement than on the anatomic structures involved

Both iritis and iridocyclitis have often been attributed to *chronic focal infection* However there are but a few patients who have the disease in whom focal infections can be demonstrated Abnormal intestinal flora and intestinal toxins cannot be demonstrated Iridocyclitis may be a part of (1) Syphilitic iritis or iridocyclitis which occurs during the secondary stage of syphilis (2) Gonorrheal iritis or iridocyclitis which occurs in a few of those who have gonorrheal urethritis (3) Tuberculous iritis or iridocyclitis which occasionally occurs (4) Iritis or iridocyclitis which may be mentioned as a complication of diabetes gout and rheumatism

Despite the fact that all are agreed that usually an iritis or an iridocyclitis is of systemic origin general examinations often do not disclose a definite cause

The chronic or relapsing form of iridocyclitis which often affects younger persons frequently is termed uveitis

The symptoms of iridocyclitis consist of pain photophobia lacrimation and interference with vision The pain is often severe and may be referred to the region of the eyeball or it may extend to the forehead or temple it is worse at night than by day Often there is tenderness of the eyeball On examination the iris appears swol

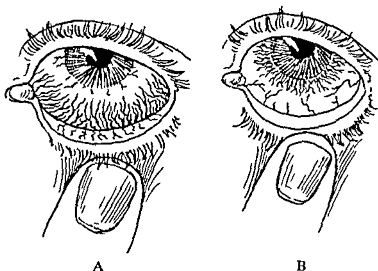


Fig 4 6 Distribution of dilated blood vessels in conjunctivitis and in iridocyclitis A conjunctivitis B iridocyclitis

len and dull it has lost its color and is now greenish blue or gray and may be of a smoky appearance The pupil is contracted irregular and sluggish in action One of the most characteristic of the external appearances of this condition is a marked circumcorneal injection and more or less conjunctival injection It is important that iridocyclitis be differentiated from an acute catarrhal conjunctivitis because in iridocyclitis the instillation of atropine is most important in treatment (Fig 4 6) Likewise the condition should be differentiated from glaucoma because the instillation of atropine in a glaucomatous eye is disastrous It should be remembered that generally in iritis the pupil is small gray and sluggish and irregular in outline after the use of atropine In glaucoma the pupil is dilated oval and immovable The cornea in iritis is transparent the cornea in glaucoma is insensitive In acute iritis the conjunctiva is usually transparent whereas in glaucoma the conjunctiva is congested and chemotic In iritis the eyeball tension is usually normal in glaucoma it is increased

Diseases of the Retina On the interior of the eye the expansion of the optic nerve forms the retina The retina is divided into two lateral halves each supplied by a cor

Pathologic Conditions There are three varieties of paralysis of the iris or iridoplegia with pupils small or of medium size as follows (1) loss of response to *light*—reflex iridoplegia (2) loss of response to *accommodation*—accommodative iridoplegia (3) loss of response to *irritation of the skin of the neck*—absent skin reflex

Common Abnormalities of Iris and Pupil The common abnormalities of the iris and pupil together with their possible diagnostic indications may be stated as follows

Iris and Pupillary Outline Inflammation of the iris is often of idiopathic origin If iritis is present the iris if normally blue or gray becomes darker the pupil responds sluggishly to the light and if the disease is unilateral is smaller than the other pupil because of the iritic congestion and consequent swelling There is a narrow zone of pinkish hyperemia around the edge of the cornea If the iris is brown or black little change of color occurs While one iris may normally differ in color from the other if one is green and the other blue the existence of iritis should be strongly suspected In many cases iritis is unilateral and is prone to relapse Syphilitic iritis occurs only during the secondary stage of the disease is commonly bilateral and rarely relapses

An irregular outline of the pupil is due to iritis and subsequent adhesions between the posterior surface of the iris and the anterior surface of the lens posterior synechia

Anisocoria Inequality of the pupils may be present in healthy persons and in those whose eyes refract unequally

Adie s Syndrome Adie s syndrome is an anomaly in which there is one myotonic pupil often associated with absent tendon reflexes The condition is generally unilateral and the defective pupil is larger than its fellow The cause is unknown except in instances where it is associated with the Pancoast syndrome

Hippus A rapid contraction and dilatation of the pupil on sudden exposure to light is normal in some healthy persons It is also seen in the early stage of acute meningitis in disseminated sclerosis and in hysteria In Cheyne Stokes respiration the pupils may be dilated during the period of rapid breathing and contracted in the apneic interval

Argyll Robertson Pupil If the pupil does not respond to light and shade but does respond to accommodation it is of the type called the Argyll Robertson pupil The condition most frequently is present as a symptom of locomotor ataxia and perhaps is due to changes in the third nerve nucleus which presides over the light reflex the portion which regulates the accommodative contraction not being involved The loss of the light reflex may also be due to degeneration of the ciliary ganglion The pupil in this condition is usually contracted because the lesions in the cervical segment of the spinal cord interfere with the dilating mechanism Late in the disease the accommodation reflex also may be lost A similar pupillary condition is seen as a symptom of intracranial syphilis and progressive paresis of the insane

Accommodative Iridoplegia With Preserved Light Reflex This condition is the opposite of the Argyll Robertson pupil It is of infrequent occurrence and may denote a lesion affecting a portion of the oculomotor nucleus in which lie the cells causing accommodative reaction It may also but not always exist with the cycloplegia which occurs after diphtheria

Unilateral Dilatation One pupil may be dilated as the result of disease of the optic nerve as in optic atrophy or there may be lessened transparency of the media as in cataract and glaucoma If the pupil of the diseased eye doe not react directly to light but does react when the light is thrown into the sound eye (consensual or indirect reaction) it shows that the disease is in the optic nerve or tract and that the oculomotor nerve of the diseased side its nucleus and nuclear connections with

complete blindness In the beginning the field of vision is usually contracted peripherally for colors and there may be reversal of the color field The blind spot is enlarged and hemianopsia or scotomas may be present

Examination for disorders of the retinas is performed by means of the ophthalmoscope and direct retinoscopy Retinitis is almost always bilateral and does not present external signs or symptoms Examination reveals an indistinctness of retinal details congestion of the disk with blurring of its edges A retinitis is characterized by circumscribed exudations appearing as soft white spots or patches of variable size which may be discrete or confluent The spots occur principally along the retinal vessels and in the macular region Often there are tortuosity and distention of the retinal veins which appear to be darker than they are when normal The vessels may be obscured in part by swelling and exudation Hemorrhages are present and are often described as rounded soft or flame shaped In retinitis accompanied by choroiditis opacities in the vitreous may be seen

Retinal Hemorrhage The vascular changes accompanying arterial hypertension are the commonest cause of retinal hemorrhages as described in the section on hypertension Chronic nephritis may be the causative condition and so also may the hemorrhagic blood states as in scurvy hemophilia purpura and the grave anemias Retinal extravasations may also be significant of malarial fever septicemia pyemia and leukemia although in the last a retinitis is commoner than retinal hemorrhage

Detachment of Retina The retina may become detached When it is detached there is a loss of vision in the visual field of this part of the retina A small loss of vision from a detachment of the retina may not be noticed by the patient However if there is an extensive detachment of the retina the patient observes a visual disturbance which may be described as a cloud or curtain hung before the eye Early symptoms are *metamorphopsia* and flashes of light or photopsia Detachments of the retina are easily visualized by means of the ophthalmoscope They are commoner in myopic eyes than in other types of eye

THE PUPIL

The pupil is the aperture in the iris which controls the amount of light reaching the lens and consequently the interior of the eye The points to be observed about the pupil are its state of dilatation or contraction inequality (anisocoria) response to light response to accommodation and the skin reflex

The responsiveness of the pupil to light is determined for each eye separately by the use of an artificial light or by asking the patient to look at the light from a window With the patient still keeping the eyes in the same direction the hands of the physician are placed over and close to but not touching the eyes One hand is then suddenly removed and the behavior of the pupil is noted Normally the pupil contracts on exposure to light—the *direct reflex* In normal eyes the pupil of the covered eye will also contract—the *indirect reflex* Particular care in this examination is desirable Artificial light in a dark room should be employed In the modern lighted room the light reflex cannot be determined with the lights on Turn the lights off before attempting to test for the light reflex with an artificial light While the patient with one eye covered looks toward the darkest part of the room the light is brought suddenly before the uncovered eye at a distance of 3 or 4 feet (about 91 or 122 cm) so that the pupillary accommodation reflex may be eliminated

Response to Accommodation Since the pupil grows smaller when the eyes converge and the ciliary muscle contracts in the act of accommodation for near objects this function should also be examined Let the examiner place his finger tip 12 or 15 inches (30 or 38 cm) from the eye nearly in line with some distant object and direct the patient to look first at the finger and then at the distant object until it is decided whether or not the pupil contracts when the patient is looking at the finger

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the corpora quadrigemina and through the latter with the opposite optic tract are not involved Unilateral dilatation may be due to irritation of the cervical sympathetic by a tumor of the neck, by aneurysm of the aorta, and when limited to the right eye by an aneurysm of the innominate artery One pupil is dilated in unilateral paralysis of a third nerve

Bilateral Dilatation Double atrophy amaurosis or blindness from any cause will obviously give rise to dilatation and failure of the light reflex Mydriasis may result from syncope shock nausea and aortic regurgitation The pupils may be dilated in neurasthenia hysteria and strong emotion in severe dyspnea and asphyxia in cerebral hemorrhage thrombosis tumor and abscess in the later stages of meningitis and diphtheritic paralysis, from lesions of the oculomotor nerve or nucleus in coma following an epileptic convulsion and after administration of drugs Some of the drugs which have a mydriatic action local or constitutional are alcohol atropine chloral chloroform cocaine hyoscyamus nitrous oxide strychnine and tobacco

When both pupils are dilated the eye has a peculiarly brilliant expression which may be particularly noticeable in mania in fevers attended with active delirium in exophthalmic goiter and in poisoning with belladonna

Contracted Pupils Contraction of the pupils (miosis) may be due to irritation of the oculomotor system to paralysis of the dilators or to the administration of drugs

The pressure of a mediastinal tumor if sufficiently great to paralyze the sympathetic (dilator) fibers in the upper part of the thorax may explain unilateral miosis Unilateral cerebral lesions so placed and of such a character as to irritate the oculomotor center may cause unsymmetric miosis

A certain degree of miosis and sluggish contraction of the pupil is normal in old age The pupils are contracted during sleep in a healthy person

Congestion of the iris such as occurs in the injected eyes in rickettsial fevers causes miosis so also with mitral insufficiency and other conditions producing venous congestion The pupils are contracted in retinitis and when photophobia exists

Bilateral disease of the cervical segments of the spinal cord and spinal column by paralyzing the sympathetic dilator fibers may allow miosis Symmetric miosis may thus be significant of locomotor ataxia in particular disseminated sclerosis general paresis spinal meningitis tumor or hemorrhage of the cord bulbar paralysis hemorrhage into the pons and tuberculous disease of the cervical vertebrae Irritating lesions of the brain—notably cerebral meningitis cerebral or dural hemorrhage and tumor or abscess of the brain—by stimulating the third nerve center will cause miosis Miosis is present in the early stage of sunstroke and may also be present in the uremic state

Certain drugs have a characteristic action in contracting the pupil particularly opium and its alkaloids Others are eserine and pilocarpine

Absent Skin Reflex If the pupil does not dilate when the skin of the neck is pinched or pricked it may be due to the atrophy of the iris in a glaucomatous eye or to posterior synechia following iritis It is not infrequent in general paresis

DISEASES OF THE LENS

The alteration of the refractive power of the lens is accomplished by changes in diameter and in shape of its anterior surface according to the distance between the object and the eye and the consequent changing of the incident angle of the light rays This alteration in the refractive power is known as accommodation

The lens is not the subject of inflammatory changes Degenerative changes in the lens invariably result in loss of transparency in the parts affected This condition of partial or complete loss of transparency is called cataract

Cataract The earliest stage in the development of cataract is an accumulation of fluid either as droplets beneath the capsule or in spindle shaped spaces between the lens fibers Clinically this stage can be recognized by inequalities in the refractive indices of the fibers and fluid which give rise to light and dark streaks when light is thrown into the eye by the mirror

The essential and common factor in the production of cataract is the coagulation of the proteins of the lens which accompanies advancing years However many different factors such as radiant heat luminous and ultraviolet rays and radium may cause cataracts Ultraviolet rays alter the permeability of the lens capsule Dinitrophenol used for reducing body weight may produce posterior cortical cataract The occurrence of cataract in tetany due to parathyroid deficiency suggests a definite association with the role of calcium metabolism Cataract also occurs in association with myotonia atrophica and ergotism

Cataracts are classified according to the situation and the extent of the opacity or opacities in the lens and according to the age of the patient

Cataracts of Congenital Origin The commonest forms of congenital cataracts are lamellar and anterior capsular less common are the central or total

Lamellar cataract occurs early in infancy The opacity in lamellar cataract is situated in the layers surrounding the center of the lens which itself usually contains punctate opacities the superficial cortex is clear Lamellar cataract is due to a period of malnutrition at some stage of late intra uterine or early infantile life Epithelial structures are most affected and characteristically epithelium persists where ordinarily it is cast off Epithelium persists in the lens and in the enamel of the teeth Lamellar cataract is almost invariably accompanied by defective enamel in certain of the permanent teeth The hypoplasia differs essentially from the condition of the teeth in congenital syphilis It is often due to maternal rubella

Cataracts may result from premature birth

Anterior and posterior cataracts are of congenital or acquired origin Anterior cataracts are designated anterior capsular and anterior cortical The posterior cataracts are posterior opacities called posterior polar and posterior cortical cataracts

The child is brought for examination on account of defective vision Objects are held very close to the eyes

Senile Cataract A senile cataract rarely occurs in persons less than 50 years of age Cataract occurs equally in men and women It is usually bilateral but develops earlier in one eye than the other Cases of hereditary predisposition have been recorded and in some of these the cataract develops at an earlier age in successive generations

The appearance of black spots in front of the eyes is usually the first symptom The spots differ from the ordinary muscae volitantes occasionally complained of in normal eyes and much exaggerated in cyclitis in that they are stationary retaining their relative position in the field of vision in different positions of the eye There may be doubling or tripling or even more duplication of the objects seen This is often worse on looking at bright lights Eventually the central area becomes affected and vision steadily diminishes until only perception of light remains

The pupil in old people is seldom so black as in the young and is sometimes distinctly gray If the grayness is uniform cataract is not diagnosed unless definite opacity is shown on examination with the ophthalmoscope This grayness without opacity is caused by increase in the refractive index of the cortex of the lens in old people and is due to increase of reflection and scattering of light

The nucleus in the common senile cataract undergoes little change and does not become opaque but eventually the whole of the cortex is cataractous The cataract is then said to be ripe or *mature* At this stage it will be found that the anterior chamber has regained its normal depth The watery fluid has been absorbed from the lens which has again returned to its normal volume This is an important guide to the most favorable time for operation

In *incipient* senile cataract radial spokes or sectors of opacity are seen with clear areas between them They are difficult to see in daylight or by oblique illumination and cataract cannot be diagnosed without confirmation with the ophthalmoscope With the pupil undilated only the ends of the spokes are seen, but when the pupil is dilated the linear opacities are often found to be the apices of sectors with their bases toward the periphery They generally begin in the lower part of the lens especially the lower nasal quadrant

Secondary Cataract A form of secondary cataract is the opacity which persists or follows after the extraction of the lens

Cataract in diabetic persons is by no means always diabetic in the proper sense of the term Senile cataract of the usual type following the usual course often occurs in elderly diabetic patients Cataract in young diabetic patients is a secondary or complicated cataract and termed *diabetic cataract*

Owing to exposure to roentgen rays or radium cataracts may form *Irradiation cataract* resembles the early posterior cortical stage of *glassworkers cataract* One course of deep roentgen rays may suffice There is a considerable latent period which may be at least two years Only the gamma rays of radium seem to be nocuous

THE VITREOUS BODY

Opacities Black specks floating before the eyes may be seen by normal persons These specks (muscae volitantes) are opacities of various kinds which cast shadows on the retina thus appearing as dark spots in the field of vision The corpuscles circulating in the retinal blood vessels at times may be seen as golden colored specks Other muscae may be due to small specks in the vitreous Some types of muscae cannot be seen with aid of the ophthalmoscope

Under abnormal conditions muscae may be of sufficient numbers to interfere with vision and to become visible by the ophthalmoscope They then indicate some disease of the uveal tract particularly of the ciliary body they are found in cyclitis and choroiditis In the more severe instances muscae may resemble flakes and threads Such large opacities are often present after hemorrhage into the vitreous

Fluid Vitreous and Specks Before the Eyes Fluid vitreous is common in cases in which opacities are present and is associated with the same causes The degeneration of the vitreous which leads to fluidity often causes the deposition of crystals of cholesterol which sink to the bottom of the vitreous chamber but are stirred up by movements of the eye They then appear as a shower of specks of a bright yellow color (synchysis scintillans)

Blood Hemorrhage into the vitreous may result from arteriosclerosis or inflammation of the retina from contusions or wounds of the eye or may appear without apparent cause Hemorrhages filling the vitreous with blood are suspected when no reflex can be obtained on throwing in light with the mirror It may then be possible to see a red mass behind the lens by oblique illumination

Severe vitreous hemorrhage may occur in apparently healthy young adults It has a great tendency to attack both eyes in succession and to recur so that though absorption may be complete in the early attacks permanent defect or complete loss of sight retinitis proliferans or dense vitreous opacities may ultimately follow from damage to the retina It is characteristic of vitreous hemorrhage that absorption usually takes place without organization owing to the absence of fibroblasts in the vitreous When organization occurs as in so called retinitis proliferans it is most marked near the disk from which membranes and strands stretch forward Even then organization rarely occurs in the absence of some general disease

Fibrous Tissue Fibrous tissue is present in the vitreous in cases of plastic iridocyclitis usually in the form of membranes stretching across behind the lens In some cases of this type especially in syphilitic and possibly tuberculous subjects minute bunches of newly formed blood vessels project from the disk or retina

Parasites Cysticercus when present may be seen ophthalmoscopically as a pearly translucent mass with peristaltic movements

THE OPTIC NERVE

As the optic nerve enters the eye it is contracted and forms the disk or papilla It is readily seen with the ophthalmoscope as a round spot somewhat lighter in color than the surrounding eyeground Coming from a depression or cup in the disk called the porus opticus are the retinal arteries and veins A certain amount of cupping is normal but if wide and deep with overhanging edges over which the vessels can be seen to dip it is indicative of glaucoma

Sometimes as in brain tumor and in malignant hypertension the papilla disk or optic nerve head is swollen When this condition occurs it is called choked disk or papilledema

Inflammations of the optic nerve known as optic neuritis consist of two groups of affections one arising within the eyeball (papillitis) and the other arising behind the eyeball (retrobulbar neuritis)

Papilledema or Choked Disk Papilledema or choked disk is a noninflammatory swelling of the optic nerve head which often results from increased intracranial pressure from malignant hypertension or from obstruction of the orbital veins The condition is usually bilateral though there may be different degrees of swelling in the two eyes In rare instances only one eye is affected

On ophthalmoscopic examination of the fundus in instances of intracranial hypertension there are swelling and protrusion of the disks distortion and tortuosity of the retinal veins and hemorrhages on or near the swollen papilla There usually is a sharp limitation of the condition to the optic nerve and to the retina immediately adjacent to the nerve head The degree of projection of the disk is determined by the difference in refraction between its most protruding part and the adjacent but unaffected retina This difference is expressed in diopters The disk is not considered to be choked unless there is a refractive difference of 2 diopters or more In the early stages of a choked disk the blind spot is enlarged As the intracranial pressure increases the vision may become gradually less until finally blindness ensues However immediate relief of intracranial pressure prior to blindness often restores vision Choked disk may occur as the result of a number of intracranial conditions such as hydrocephalus brain abscess and suppurative meningitis It is important to keep in mind that choked disk accompanied with neuroretinitis is characteristic of malignant hypertension

Choked disk does not accompany growths in the pituitary body It occurs late if at all in lesions of the motor cortex and is most frequent in neoplasms of the cerebellar parieto occipital regions and midbrain The condition is symptomatic of tuberculous nontuberculous and suppurative forms of the meningitis

Retrobulbar Neuritis Inflammations of the optic nerve behind the eye are termed retrobulbar neuritis There is slight or no change in the appearance of the disk unless a tumor of the brain is present

The common cause of retrobulbar optic neuritis is syphilis In rare instances optic neuritis may be caused by encephalitis meningitis acute febrile diseases or by poisoning especially lead Retrobulbar optic neuritis may develop in young adults in association with disseminated sclerosis without other symptoms Diphtheria and diabetes may cause this disorder of the optic nerve Unilateral optic neuritis occurs only when there is disease of the bones of the orbit or inflammation of the orbital tissues It often exists without an assignable cause

The first symptom is unilateral or bilateral blurring of vision with pain in and above the affected eye brought on by ocular movements The reduction of vision is rapid reading is impossible within 2 or 3 days after the onset One eye may be affected first but the condition is bilateral

The pupils are equal and circular and react equally to light They are sluggish Central vision is reduced or abolished The visual field except for the central scotoma is normal The optic disk at first is congested and shows haziness of the margins Temporal pallor then appears which bears no relation to the disturbances of vision *Usually within a few weeks vision begins to improve Rarely is vision lost* in the first attack There is a strong tendency for the trouble to recur

Retrobulbar neuritis without edema of the disks is distinguished from that due to various poisons such as lead arsenic thallium and methyl alcohol as indicated by the history and appropriate examinations

Neurosyphilis is not of etiologic importance Tumors of the hypophysis pressing on the intracranial portion of the optic nerve can be excluded on the basis of roentgenograms of the sella turcica

The prognosis of retrobulbar neuritis is at best doubtful for complete recovery or for complete blindness Disseminated sclerosis may appear immediately or after many years When disseminated sclerosis develops in other parts of the nervous system the process is acute and severe

Toxic Amblyopia There are a number of causes for toxic amblyopia among which quinine methyl alcohol overuse of tobacco vitamin deficiency and uremia may be mentioned

The toxic amblyopias are manifested by a gradual diminution in acuteness of sight The dimness of vision may be more definite in bright light than in dim light There are central color scotomata The ophthalmoscopic signs are usually minimal but pallor of the temporal side of the disk color scotoma may occur as the condi tion progresses

Atrophy of the Optic Nerve Inflammation of the optic nerve with congestion hemorrhage and exudation into its substance may affect the nerve posterior to the globe *retrobulbar neuritis* the nerve at its end *papillitis* the retina *retinitis* In course of time there may be connective tissue proliferation and consequent atrophy of the nerve fibers giving rise to secondary *optic atrophy*

Atrophy of the optic nerve occurs also as simple atrophy or primary atrophy which is a noninflammatory condition The cause is unknown There may be sec ondary atrophy of the optic nerve which is postneuritic secondary inflammatory atrophy or atrophy of nutritional origin

Primary Atrophy In primary or simple optic atrophy there is degeneration of the nerve fibers The common causes of simple atrophy are cerebrospinal diseases *especially tabes dorsalis during its inception Simple atrophy is less frequent in* general paralysis of the insane It may occur in multiple sclerosis When it does occur as a part of multiple sclerosis it follows retrobulbar neuritis it rarely results in complete blindness though often there is a transient blindness Simple optic atrophy may accompany systemic syphilis malaria diabetes acromegalia oxy cephaly profound hemorrhage with loss of consciousness (anoxia) the specific fevers arteriosclerosis and certain poisonings including methyl alcohol lead and arsenic poisoning It results from a *thrombosis of the central retinal arteries* and it may occur in connection with choroiditis retinitis glaucoma aneurysm of the in ternal carotid artery and orbital inflammation There is a hereditary form of the disease which is commoner in young men than in others

Secondary Atrophy Secondary optic atrophy is usually consequent to an optic neuritis expanding intracranial lesions and tumors of the optic nerve Retrobulbar neuritis syphilitic chorioretinitis and retinitis pigmentosa may also be responsible for atrophy In some who have chronic hydrocephalus a greatly distended third ventricle may press on the chiasm and produce optic neuritis and atrophy

Optic Atrophy and Arteritis An optic atrophy may result from an arteritis as in periarteritis nodosa or in the condition known as temporal arteritis

Syphilitic Optic Atrophy Bruetsch observed the existence of both the inflammatory and the degenerative phases of syphilitic optic nerve atrophy He emphasized that the degenerative changes in optic atrophy are preceded by an inflammatory exudative phase Visual field defects may be caused by the exudative processes in the marginal zone before atrophy of the nerve fibers has taken place and such defects may improve or disappear after antisyphilitic treatment Klauder and Meyer have stressed the necessity of an early ophthalmologic and perimetric examination of all patients who have neurosyphilis regardless of whether the serologic reactions of the blood and spinal fluid are positive or negative The presence of pupillary abnormalities calls for examination of the cerebrospinal fluid in order to exclude optic nerve atrophy It appears therefore that the problem of syphilitic optic nerve atrophy as a cause of blindness is concerned principally with early diagnosis The ophthalmoscope and perimeter are as essential as the spinal puncture in the diagnosis and management of syphilis and its complication primary optic nerve atrophy

In optic atrophy from any cause the patient complains of a profound disturbance of vision consisting of on examination a contraction of the visual fields blind spot enlarged color fields reversed and scotoma

INJURIES TO THE EYE FROM CONTUSIONS AND RADIANT ENERGY

There is no part of the eye which may not be injured so seriously by contusion or radiant energy as to diminish vision In some cases the changes are progressive In all cases a guarded prognosis should be given when these injuries have been sustained

Contusions A contusion may vary in severity from a small abrasion of the cornea to rupture of the eyeball

Cornea Injuries to the cornea incident to sharp objects manifested by only a slight abrasion revealed by staining with fluorescein heal immediately A similar small abrasion of the cornea incurred by a sudden fall or a stroke from a blunt instrument causes severe pain The pain is increased on moving the lids there is much lacrimation as well as reflex blepharospasm The sequence of events a fall slight corneal abrasion and severe pain is originated by a pre-existing corneal ulcer or results from injury to the fifth cranial nerve at the time of the fall or from pre-existing disease of the fifth cranial nerve

In some instances of injury to the fifth cranial nerve the corneal abrasion may heal quickly However after days weeks or even months acute pain and lacrimation commence These symptoms are more evident on first opening the eyes in the morning than at other times There is no doubt that in these cases the epithelium is abnormally loosely attached to Bowman s membrane and is likely to be torn off by the lid on waking Such looseness of epithelium and formation of vesicles are characteristic of lesions of the fifth nerve and it is probable that recurrent erosion is due to this cause

On examination deep opacity may be found which is due to accumulation of lymph in the interlamellar spaces These opacities are generally transient and clear up

Examination by means of the slit lamp reveals that all contusions of the eyeball are followed by the deposition of uveal pigment on the posterior surface of the cornea There may be ruptures in Descemet s membrane owing to its elasticity but ruptures of the cornea are rare

Sclera Rupture of the globe is generally due to its being suddenly and violently forced against the orbital walls It is often due to falls or to the eye being forcefully struck by a projecting object the force having been applied from the inferolateral direction where the eyeball is least protected by the orbital margin The wound is oblique being farther forward internally than externally It is situated concentrically with the corneal margin The conjunctiva is usually left intact but severe injuries

to other parts of the eye are inevitably present The iris is often prolapsed or torn away (iridodialysis) or retroflexed The lens may be expelled from the eye or may escape under the conjunctiva or be forced back into the vitreous A displaced lens leaves the anterior chamber deep The anterior chamber contains blood and there may be hemorrhage into the vitreous Detachment of the retina may occur with or without subretinal or subchoroidal hemorrhage The eye shrinks and sight is lost

Iris Most injuries to the iris caused by contusion are due to direct blows with a sudden inward displacement of the cornea

Traumatic mydriasis may follow a contusion The pupil is immobile, and remains moderately dilated and paralyzed to accommodation permanently Radiating lacerations of the iris sometimes extending to the ciliary margin may occur In iridodialysis a black biconvex area is seen at the periphery and the pupillary edge bulges inward With the ophthalmoscopic mirror a reflex can be obtained through the peripheral gap Iridodialysis is permanent Apart from other injury however it rarely causes serious disability

Lens Injuries to the iris may cause rupture of the suspensory ligament The lens is forced back into the vitreous With the pupil dilated the edge of the lens may be seen as a gray convex line by oblique illumination but more readily as a black line with the ophthalmoscopic mirror The lack of support to the iris is detected by its unsteadiness revealed on movement of the eye

In partial rupture of the suspensory ligament there is astigmatism which is much increased on movement of the head as the lens is thus tilted Total dislocation of the lens into the vitreous causes a deterioration of vision

In a lens displaced so much laterally that the edge crosses the pupil uniocular diplopia is present In subluxation the lens is likely to become opaque owing to malnutrition The pressure of the edge of the lens on the back of the iris and on the ciliary body often originates a severe iridocyclitis and endangers the other eye by sympathetic ophthalmia Secondary glaucoma is frequent

With or without dislocation of the lens concussion cataract occasionally follows a contusion In most of these instances the capsule has been ruptured The lens gradually becomes opaque The cataract spreads throughout the cortex until the appearance of a mature cataract is present

Vitreous Hemorrhage into the vitreous is common after contusion The vitreous chamber may be filled with blood and the eye is without reflexes On oblique illumination a dull red hue may be seen The blood may become almost completely absorbed but cloudy opacities remain

Choroid Rupture of the choroid occurs as the result of severe contusion It has also been caused by a bullet passing through the orbit behind the eye Immediately after the injury the view is obscured by extravasation of blood When the blood has become absorbed the rupture appears as a curved white streak over which the retinal vessels pass If the choroid is ruptured near the macula there is loss of central vision Ruptures of the choroid in which the macula is not involved cause little impairment of vision

Retina Detachment of the retina may be due to contusion However spontaneous detachment of the retina often occurs

Blows on the eye may change the normal bright red color of the retina to a light gray near the papilla This change in color which is probably due to edema disappears after some days and vision is usually restored to normal In some instances vision may be good at first but then central vision diminishes and pigmentary deposits develop at the macula Hence prognosis should be guarded in all cases of serious blows on the eye

Optic Nerve The optic nerve is frequently injured in fractures of the base of the skull Injuries by sticks or knives penetrating the orbit are rare Avulsion of the

optic nerve is extremely rare but occurs as the result of wounds from flying missiles or bullets

Radiant Energy Radiant energy may be divided into two types (1) the electro magnetic radiation and (2) the corpuscular radiation consisting of subatomic particles the most noteworthy of which are the electrons (variously called beta rays and cathode rays depending on their source) and the neutrons (Cogan)

The radio and diathermy waves cause no effect on the eyes except in the immediate vicinity of a transmitter An electric current consisting of beamed movements of electrons may cause ocular injury or other injury through electrolysis or by the production of heat in the tissue

The long infrared radiation which may give rise to cataracts is largely derived from open furnaces used by professional glass blowers and foundrymen Infrared cataracts occur only after exposure to intense heat for many years The cataracts resulting from infrared radiation are characteristically posterior cortical forming at first saucer like opacities belonging to the general group of so-called cataracta complicata

The short infrared radiation which has been harmful to the eye (retinitis) may be obtained from electric arcs the sun and quartz and mining lamps

The amount in the abiotic range of ultraviolet radiation filtering through the atmos phere from the sun is too slight to be harmful except under extraordinarily clear condi tions or at high altitudes

Keratitis resulting from ultraviolet radiation of the sun occasionally occurs with pro longed exposure at high altitudes or in the presence of large snow fields The lesion consists predominantly of punctate erosions of the epithelium in the exposed portions of the cornea The symptoms come on after a latent period of 5 to 12 hours the patient has no feeling of ill effects from being exposed to harmful radiations at the time of exposure The effects are cumulative during an interval of 24 hours and unlike the skin reaction no tolerance is acquired by repeated exposures The patient s symptoms are severe photophobia pain epiphora and blepharospasm which are incapacitating at the time but seldom lead to permanent injury of any part of the eye

The corneal lesion resulting from grenz rays and beta rays is keratitis with severe symptoms consisting of epiphora foreign body sensation and photophobia but the latent period is longer and the duration is longer than is the case with ultraviolet effects Susceptibility of the lens to roentgen rays is said to be an inverse function of age the most vulnerable being fetuses irradiated in utero

Glaucoma may follow roentgen irradiation of the eye It arises several weeks after irradiation and usually in association with uveitis

The latent period for the development of cataracts following irradiation is usually from 6 months to 2 years but may be as long as 10 or more years In general the greater the dose the shorter the latent period

A roentgen ray cataract begins characteristically as an opacity at the posterior pole of the lens It may remain stationary but is usually progressive Progression is usually indicated by the appearance of vacuoles in the anterior cortex and an anterior subcapsular haze from proliferation of the lens epithelium

Cataracts from gamma rays and neutrons are similar to those produced by roentgen rays as regards latent period and structure and similarly occur with doses that produce no other clinically evident abnormality

Radio waves are not known to produce any physiologic or pathologic effects on the eye and the effect of diathermy waves is that of a thermal coagulation when locally applied

The long or far infrared rays probably can cause a thermal burn of the retina The susceptibility to retinitis may be influenced by nutritional factors which induce a sensitization to visible radiations so that retinal lesions may result from exposure to sunlight (Cogan)

VISION

The disorders of vision which are of diagnostic importance are hemianopsia alterations in the color fields amblyopia and amaurosis

Minor Disorders If objects seen have a yellow tint it may be due to jaundice or the administration of drugs particularly of santonin In exhausted neuropathic women or children overuse of the eyes may cause everything to appear red The small, beaded, semitransparent threads or dots (muscae volitantes) seen in looking at some clear expanse of light are as stated of small diagnostic importance They appear to be most abundant in hysteria and functional disorders Flashes or small luminous points of light before the eyes occur commonly after quick changes in position coughing or straining and usually are not of diagnostic import However they may constitute the aura of epilepsy or migraine Migraine may be preceded by scotomata resembling a cloud the edges of which are brilliantly lighted or colored The scintillating scotomata appear as bright dots whirling figures and as bright lines before the eyes

Failing Vision The causes of failing vision arise from the cornea iris ciliary body and choroid retina optic nerve lens and vitreous and from disuse The following paragraphs contain a summary of the views expressed by Post in regard to failing vision

Among the corneal causes of visual failure tuberculosis is common It is usually considered an allergic manifestation Corneal degeneration is frequent in old persons The central areas anteroposteriorly may become less transparent Nodules appear in the posterior elastic membrane that look like small drops of water on the rear surface of the cornea when seen with the corneal microscope Epithelial degeneration occurs

Corneal ulcers are an important cause of visual failure They occur throughout life their causes varying from gonorrhea in infancy through phlyctenular keratitis in youth to malnutrition and the physiologic changes incident to age In preadolescence the interstitial keratitis of syphilis may cause blindness

Allergic diseases of the iris ciliary body and choroid are as frequent in ophthalmologic practice as allergic diseases of other structures are in every other department of medicine Allergic diseases of the eye are chiefly conjunctivitis and lid reactions the former the so called vernal conjunctivitis and as such they are seldom responsible for failing sight but allergic scleritis and uveitis are important causes for gradual loss of vision

In tuberculosis sections of the lesions in the uveal tract seldom reveal the tubercle bacilli

The most serious form of uveitis is sympathetic ophthalmia which is probably due to sensitivity to uveal pigment Differential diagnosis of allergic uveitis depends on the presence of the inherited allergic state the elimination of other more easily identifiable inflammations such as leprosy syphilis and sympathetic diseases and finally identification of the offending antigen by means of allergy tests

Retinal causes of diminished vision are numerous Diabetes affects the eye late in the course of the disease particularly in the form of chorioretinitis though undoubtedly some cataracts have diabetes as their cause Diabetes may be the cause of gradual loss of sight Though the onset of such loss of sight is late once it has started it is implacable its presence is to be suspected in gradual visual failure occurring in the middle years of life There is no pain and there are no external manifestations Often the first ocular symptom is visual failure and when this occurs the chorioretinitis is severe It may take place in patients in whom the diabetes is under control The usual therapy for diabetes is completely ineffective with respect to established retinal lesions

In cardiovascular renal disease both sudden and gradual visual loss occur The former is especially associated with large retinal hemorrhages and the latter with arteriosclerosis and small hemorrhages and exudates The eyegrounds reveal the changes of malignant hypertension as early as these changes are to be found anywhere in the body

Degenerative changes frequently accompany old age The retina suffers in this condition primarily from malnutrition and probably an accumulation of waste products Glaucoma causes a failure of vision

Optic atrophy is a major reason for failing sight Among its most important medical causes are syphilis hypophyseal tumor multiple sclerosis and drugs In these diseases the ophthalmologist is often the first to make the diagnosis In 8 of each 10 patients who have supratentorial neoplasms choked disks are present whereas in those who have hypophyseal tumors atrophy of the papillomacular bundle is the rule In multiple sclerosis the onset may be associated with optic neuritis but more often there is simply temporal pallor of the nerve head with loss of central vision

Failure of sight from optic atrophy may be sudden as from poisoning from drugs such as methyl alcohol (CH OH) (see Methyl Alcohol Chapter 19)

Peripheral field changes may be advanced before they attract the patient s atten tion as for instance in chronic simple glaucoma the most frequent cause of failing vision and blindness of middle and old age This disease may not cause pain or redness of the eyes and is noticeable only to a discerning patient by a diminution in the visual field or halos about lights Cataracts are common and their role in failing vision is well known

Congenital Color Blindness Congenital color blindness or congenital color amblyopia occurs in 3 or 4 per cent of boys and men and in about 0 3 per cent of girls and women The condition generally affects both eyes and is often hereditary The functions of the eyes are otherwise normal There are a number of tests for color vision which are particularly useful in the examination of employees in certain occupations in which perfect color perception is essential for example in railway steamship and aviation services as well as in motor driving In transportation services the most commonly used signals are red and green the colors in which most color blind persons are defective

There are a number of well known tests for color vision Stilling s test Ishihara s test and Holmgren s test

The Holmgren test is frequently used and is very convenient It employs a large assortment of colored worsteds This collection includes (1) test colors pale green light pink and bright red (2) match colors lighter tints and darker shades of test colors and (3) confusion colors yellow brown gray drab fawn mauve pale blue and the like The test is performed in good daylight The pale green sample is given to the individual who is asked or required to select colors that match the test sample If he does this cor rectly he has normal color sense If he not only fails to select similar colors but also confuses colors and in addition shows a certain hesitancy his color sense is defective Next a pink skein is selected and the person being examined is asked to match it If besides similar skeins he selects blue or violet he is red blind If he selects green or gray he is green blind Finally he receives the bright red test skein for matching If besides red he chooses green and brown colors darker than the red he is red blind If he selects shades of those colors lighter than the red he is green blind

Effect of Teaching on Color Blindness Some men can be taught how to correct for color blindness Educational programs for those who are color blind have been con ducted which permit them to pass tests for color blindness In one technic (Lepper) the subjects are instructed to get someone with normal color vision to assist them and to trace the shapes of the figures carefully even in those charts in which they can see the figures fairly well They are instructed to continue working on harder and harder charts Thus the person concerned builds a learning curve and with sufficient time seems to master the color manual It seems quite likely that those who hav d sturba ices of color vision do fix in their minds appearances which are associated with certain colors when viewing objects that have for them some familiarity

Alterations in the Field of Vision The alterations in the size and shape of the visual fields which may be of value in diagnosis are hemianopsia contraction of the visual fields for light and contraction of the color fields In connection with hemi

anopsia blindness due to disease of the optic nerve or its central connections is diagnostically important

The diseases which may affect the optic tract are neoplasms syphilitic meningitis gummas and tuberculous meningitis In these cases the crus may be implicated causing hemiplegia or as the result of basal lesions ocular muscle paralysis may be present The absence of any form of aphasia in right hemiplegia is against a central lesion and in favor of an affection of the optic tract

Hemianopsia A condition termed hemiopia hemianopia or hemianopsia is blindness of one half of the field of vision

Since in the majority of cases of hemianopsia there is organic disease of the brain hemiplegia and hemianesthesia on the same side with the hemianopsia are uncommonly associated symptoms, and in left side lesions aphasia If athetosis coexists with lateral hemianopsia it is significant of a lesion involving only the posterior gray mass of the optic thalamus If the hemianopsia involves a quadrant and not a semicircle or is otherwise incomplete and if mind blindness or word blindness coexists the lesion is usually cortical Dimness of vision in one eye and a marked contraction in the visual field of the other together with mind blindness (seeing but not recognizing objects) is a combination indicative of a lesion of the angular gyrus

Hemianopsia in rare instances may be due to hysteria in which case the conjunctiva is usually anesthetic and hemianesthesia is present But by far the commonest alterations of vision in hysteria are contraction of the visual fields and changes in the color sense The ophthalmologist can usually definitely distinguish the contraction of the visual fields due to hysteria from those due to organic disease

Total blindness of one eye may be due to a lesion of one optic nerve or disease of one occipital lobe and if in addition there is hemianopsia of the opposite eye it may be due to disease of the decussating fibers in the center of the chiasm together with the direct fibers of one lateral angle If there is no disease of the retina or other ocular structures sufficient to account for the loss of sight a lesion at the decussation is likely to be present Total blindness of both eyes will be caused by disease affecting the entire chiasm or bilateral lesions of the cuneus

Hemianopsia or blindness of one half of the visual field is of different varieties and together with the qualifying terms employed the name refers to the field of vision from the point of view of the patient—not to the retina Thus right temporal hemianopsia means that the outer or temporal half of the field of vision of the right eye is blind so that if the eye is fixed on a point directly in front objects to the right of the point of fixation will not be perceived Therefore remember that blindness of the temporal half of the field of vision is due to loss of function of the inner or nasal half of the retina because of the crossing of the light rays in the media and so with other varieties

If the hemianopsia affects both visual fields it is *bilateral* if corresponding halves (both right or both left) are implicated it is *homonymous* or lateral If both temporal or both inner fields are implicated it is *heteronymous* Furthermore the inner fields are termed *nasal* the outer *temporal* Very rarely the upper or lower halves of the visual field are effaced and if so the condition is then called *superior* or *inferior* hemianopsia Hemianopsia may be partial only a portion of the half field being blank The unaffected half may retain its normal dimensions or as in some cases be reduced in size

The presence of hemianopsia may be detected by those untrained in ophthalmology but delineation of these defects is a task for the ophthalmologist To detect the presence of hemianopsia each eye must be tested separately the resting eye having been covered by a card The patient being placed with his back to the light is told to fix the uncovered eye on that part of the physician s face (placed about 2 feet (61 cm) away) which is most nearly on the horizontal plane of the eye to be

tested The finger or a bit of white paper is then brought first from one side then from the other to the median line If the patient cannot see the paper until it has passed nearly or quite to the line of fixation hemianopsia exists The vertical extent of the field is tested in a similar manner by bringing the object from above downward and from below upward

The diagnostic indications of the various forms of hemianopsia are as follows

UNILATERAL NASAL HEMIANOPSIA Blindness of the nasal half of the field an extremely rare occurrence indicates a very limited lesion at the outer angle of the chiasm

BINASAL HEMIANOPSIA Also rare is blindness of the nasal half of both fields requiring for its manifestation two symmetric lesions of both outer angles of the chiasm or the outer sides of both optic nerves

BITEMPORAL HEMIANOPSIA Blindness of the temporal halves of both fields is produced by a lesion involving the anterior angle or central portion of the chiasm

The disease (affecting the chiasm) which is most frequently responsible for the three preceding varieties of hemianopsia is tumor or enlargement of the pituitary body with which may be associated diabetes insipidus proptosis and a flow of fluid from the nostril Other causes are tubercles tumors cysts basilar meningitis periostitis exostosis fracture of the body of the sphenoid bone and gumma The last may give rise to evanescent recurring hemianopsia When the chiasm is affected the lesion is likely to be progressive and gradually involves both crossed and uncrossed fibers thus causing total blindness of one or both eyes according to the extent of the destruction Lesions of the chiasm may be attended by symptoms indicating involvement of the olfactory and trigeminal nerves and of the nerves of the ocular muscles namely anosmia anesthesia of conjunctiva and cornea and ocular paralysis

RIGHT OR LEFT LATERAL (HOMONYMOUS) HEMIANOPSIA Blindness of the right or left half of both fields is significant of a lesion situated at some point between the chiasm and the cortical center in the occipital lobe namely the optic tract the pulvinar or posterior gray mass of the thalamus the anterior corpora quadrigemina the fibers passing from the thalamus and corpora to the occipital lobe either in the internal capsule or the optic radiations or the cortical center itself

Contraction of the Visual Fields for Light In order to determine the presence of this symptom the examiner invokes the aid of an ophthalmologist and his perimeter as it cannot be ascertained by any less accurate test Leaving out of consideration glaucoma optic atrophy or other organic disease by far the most important indications of a nearly uniform contraction of the visual fields relate to the presence of hysteria and traumatic neuroses As a rule one field presents a greater contraction than the other In neurasthenia while there may not be a true concentric limitation of the visual field yet the fatigue of continued testing will sometimes develop a decided temporary diminution in its size

Contraction of the Visual Fields for Color The color fields are tested by the perimeter using bits of colored paper Passing from the circumference to the center of the field the perception of white objects is found to have the widest extent A little nearer blue is perceived then red then green

Limitation of the color fields is a common symptom in hysteria and traumatic neuroses Sometimes a transposition of the normal fields for different colors will occur Like vision in general the color perception may be much reduced or abolished by optic neuritis or atrophy

Amblyopia and Amaurosis *Amblyopia* is a reduction in the acuteness of vision a dimness or partial blindness and cannot be relieved by glasses The condition is not dependent on any visible changes in the eye *Amaurosis* is a total loss of sight without appreciable ocular lesions The causes producing these forms of partial or total blindness are as a rule of a functional character affecting either the centers or the retinas or if organic not producing noticeable alterations in the appearance of the retinas The loss of vision is usually sudden bilateral and in most cases temporary

Obstruction or thrombosis of the central artery causes a sudden loss of vision commencing at the circumference of the visual field and extending to the center It

is frequently indicative of cardiac or vascular disease namely, bacterial endocarditis valvular disease particularly mitral stenosis atheroma of the large arteries, aneurysm of the aorta and thrombosis of the pulmonary vein very rarely it occurs in rheumatic fever and chorea In a very small proportion of cases the sudden loss of sight in chronic nephritis is said to be due to this accident or to separation of the retina

The most frequent causes are uremia diabetes and certain drugs large hemorrhages migraine and hysteria When occurring in uremia loss of sight ordinarily follows coma or convulsions is sudden and usually lasts but 1 or 2 days Quinine and the salicylates may cause sudden amaurosis and it sometimes occurs in alcoholism Severe hemorrhages particularly those from the stomach may be responsible for a sudden failure of sight Migraine may exhibit as one of its symptoms a fugitive blindness perhaps of only one half the visual field (hemianopsia) The amaurosis of hysteria is associated with emotional states and other characteristic symptoms and no matter how complete the apparent loss of sight may be the pupillary reflex both direct and consensual is retained

Hysterical amblyopia usually affects young girls and women and occasionally boys and young men This form of amblyopia is most often bilateral but it may be unilateral The most constant symptom is reduction in vision amounting to complete blindness The field of vision is contracted concentrically both for white and for colors The pupillary reflexes and ophthalmologic examinations reveal no abnormalities

Simulated amblyopia is a resort of malingerers who frequently pretend various degrees of loss of vision in one or both eyes in attempts to recover damages for lesser injury The tests for simulated amblyopia are difficult to perform because the claimant often has been coached in the various devices of evasion such as closing one eye and then the other during the test in order to ascertain which eye is being examined The ophthalmologist is aware of all of these difficulties

Blindness in both eyes is more difficult to detect than blindness in one eye Contraction of the pupil to light is presumptive evidence of some sight but in rare instances the pupils react to light in total blindness In malingering when a claim of blindness is made the ophthalmologist by the use of special tests with lights and prisms can detect evidences of eyesight

Night blindness or *nyctalopia* is a condition in which the sight is good by day but deficient at night Sometimes there is a history of exposure to bright light It may accompany certain forms of atrophy of the optic nerve such as that manifested by retinitis pigmentosa Night blindness may result from a deficiency of vitamin A Xerophthalmia is often present at the same time

Hemeralopia is a condition in which the patient sees better in an obscure light than in bright sunlight

Ocular Manifestations of Malaria Malarial amblyopia occurs as a result of the action of the malarial toxin on the optic nerve and retina Often optic neuritis and papillary edema result from the blocking of the retinal and choroidal vessels by parasites and leukocytes The retinal hemorrhages are usually small multiple and peripheral large macular hemorrhages do occur in the malignant types of malaria Ulceration of the cornea is a common ocular sequela of malaria and recurrent iritis is frequently associated with this keratitis The complaints are loss of vision frequent headaches in the occipital and temporal regions dizziness pain in the eyeballs tenderness on palpation photophobia lacrimation and spots before the eyes

The objective findings are irregularity of the pupils retinitis of the atrophic type usually in or around the macular area unusual concentrations of choroidal pigmentation generalized hyperemia of the retina and nerve head optic neuritis both mild and severe optic atrophy and in a few cases a severe progressive chorioretinitis and uveitis

Refraction and Errors of Refraction In the normal eye light passes through media of different densities being refracted at the surfaces of junction of the media so

that the incident ray comes to focus on the retina This condition is termed emmetropia

If the incident parallel rays come to focus anterior to or before they reach the retina the condition is known as myopia or nearsightedness

In hyperopia hypermetropia or farsightedness there is a lack of refracting power sufficient to bring to focus the incident or parallel rays of light on the retina

Myopia The common cause of myopia is an abnormally long anteroposterior diameter of the eyeball which results in the condition termed axial myopia Curvature myopia results from aberrations in the curvatures of the refracting surfaces of the cornea and lens which bring the incident light rays to a premature focus Abnormal positions of the lens may create similar difficulties Myopia may arise when the refractive indices of the cornea aqueous humor or the lens are changed significantly

Myopia is rare in the newborn but its incidence increases rapidly until the age of about 20 years is attained Its incidence has no relation to any uses of the eye but excessive use of a myopic eye is deleterious The cause of myopia is some predisposition which provides the requirements for myopia to develop

In myopia a crescentic posterior staphyloma (myotic crescent) in the fundus of the eye is present and this development is seemingly of congenital origin This sort of staphyloma is associated with some distortion of the papilla

In mild or low myopia there is indistinct distant vision In more severe or high myopia there is indistinct vision and discomfort after near work as the result of disproportion between the efforts of accommodation and convergence The eyes are sensitive to light Black spots are seen floating before them and sometimes flashes of light are noticed Flashes of light before the eyes are often a symptom too of detachment of the retina In very high myopia the eyes are prominent the pupils are large and the anterior chamber appears deeper than normal probably only owing to the dilatation of the pupil There may be an apparent convergent squint A true divergent strabismus may be found either concomitant or affecting only one eye Vision may be very poor even with correction scotomas may be present both central and peripheral

The ophthalmologist usually reports normal findings on ophthalmoscopic examination of the myopic eye *In moderate myopia there is a myopic crescent If there is a* high error of refraction a posterior staphyloma is recorded often with a change in the course of the retinal vessels and degenerative changes in the choroid—a chorioretinal atrophy There may be described hemorrhages in the macular region Vitreous opacities and a black spot at the fovea may be present Detachment of the retina is likely to occur in a myopic eye

With a refractive error of up to 5 or 6 diopters myopia unless in children of preschool age has a good prognosis If there are 10 diopters or more of myopia the prognosis is not good Serious retinal damage or detachment of the retina is likely to ensue

Hypermetropia (*Hyperopia*) Hypermetropia is almost always present in the infant Its incidence decreases rapidly during the first two decades of life

In many especially the young with ample reserve powers for accommodation there are no symptoms unless the condition is extreme When the powers of accommodation are insufficient from any cause there ensues eye strain The eyes after use ache feel dry and require excessive blinking Rarely there may be lacrimation

In aging persons the far point is beyond the range of comfortable reading The print has to be held farther away than usual in order to read at all In the aged the hypermetropia is index hypermetropia and much of the deficient vision may be due to an increased refractive index at the cortex of the lens

The conjunctivae and edges of the lids become reddened Headache often frontal follows excessive use of the eyes Ophthalmoscopically the fundus may exhibit no abnormality A bright reflex is commoner in hypermetropic than in

emmetropic or myopic eyes The inferior crescent and an abnormal tortuosity of the retinal vessels are common in hypermetropic eyes In some cases there is a pseudopapillitis

Astigmatism Astigmatism is present when there is defective curvature of the refractive surfaces of the eye as a result of which a ray of light is not sharply focus d on the retina The light is spread over an area of the retina and not brought to a focus on a spot of the retina There are many varieties of astigmatism Each variety is named according to its causation by prefixing an appropriate adjective or adjectives for instance corneal astigmatism which is due to irregularity in the curvature or refracting power of the cornea

Regular astigmatism designates the condition in which the refractive power of the eye shows a uniform increase or decrease from one meridian to another being practically constant in each meridian and the image produced is regular in shape either a line or an oval or a circle

Regular astigmatism is likely to cause the worst forms of asthenopia or eye strain the asthenopia in these cases is only in part accommodative It is often worse in the lower degrees of astigmatism than in the higher Aching of the eyes severe headaches and migraine are matters for complaint The eyes become fatigued easily The letters of print run together

Anisometropia A slight anisometropia is almost the rule It is a condition in which the refraction of the two eyes shows a difference In mild forms of anisometropia binocular vision is imperfect In high grade anisometropia binocular vision can be obtained only by the use of correct lenses Distinct vision even with correction may be only uniocular and the eye which is not in use may become divergent In some instances if one eye is emmetropic the other myopic the emmetropic eye may be utilized for distant vision and the myopic eye is used for near vision

Anisometropia is usually of congenital origin The symptoms are variable and may not be noticeable to the child

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5

TOPOGRAPHY AND REGIONAL DIAGNOSIS OF DISEASES OF THE NECK

The neck by means of the cervical vertebrae and muscles elevates the head and allows it to be moved freely in different directions It is interposed between the portals of entry to the respiratory and the digestive system and the thoracic organs It is therefore subjected to secondary invasions by the agents of infectious diseases as well as the extension of malignant processes from both directions In addition the neck may be involved in disease arising in its own organs Some of the diseases are of congenital origin The majority of the congenital diseases such as branchiogenic and thyroglossal duct cysts are remnants of vestigial organs

In this portion of the text are considered the structures of the neck in the median line those regions or triangles on each side anterior to the sternocleidomastoid muscle those posterior to this muscle between it and the trapezius (Fig 5 1) and finally the air passages

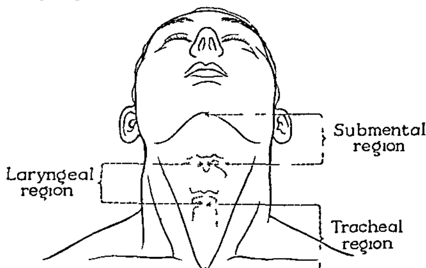


Fig 5 1 Median regions of the neck

REGIONS OF THE NECK

Median Portion Viewed from in front the median portion of the neck may be divided into three regions submental laryngeal and tracheal

The *submental region* extends from the chin to the lower border of the body of the hyoid bone it is limited laterally by the anterior belly of the digastric muscle on each side

The *laryngeal region* extends from the under surface of the hyoid bone to the lower edge of the cricoid cartilage Laterally it is limited to the space occupied by the larynx The thyroid cartilage (Adam s apple) is included in this region as a part of the larynx

The *tracheal region* extends from the lower edge of the cricoid cartilage to the top of the sternum Just above the sternum between the sternal origins of the sternocleido mastoid muscles is the suprasternal notch Laterally the region is limited by the sides of the trachea

Triangles of the Neck The ventral and lateral aspects of the neck are divided into anterior and posterior primary triangles by the sternocleidomastoid muscle The base of the anterior triangle is the border of the jaw and of the posterior triangle the clavicle The anterior cervical triangle is secondarily *divisible into three small triangles the submaxillary* between jaw and digastric muscle the *superior carotid* bounded by digastric sternocleidomastoid and omohyoid muscles and the *inferior carotid* (or muscular) *triangle* antero-inferior to the omohyoid muscle The posterior cervical triangle is divided by the inferior belly of the omohyoid muscle into the *occipital* and *omoclavicular triangles* Each of these triangles is at times further subdivisible into minor regions (Fig 5 2)

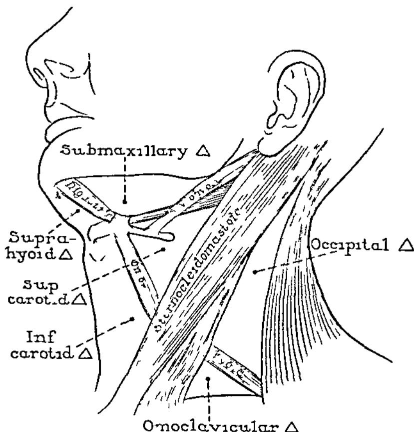


Fig 5 2 Triangles of the neck

STRUCTURES TO BE FELT IN THE MEDIAN LINE

Some necks are short and stout others are medium in length and thickness others are long and thin and there are many gradations between these types For this reason it sometimes may be easy sometimes difficult to identify the structures

On palpating downward from the symphysis of the mandible the finger discerns a hollow On pressing into this hollow the finger rests between the digastric muscles on each side and on the mylohyoid muscles beneath Still deeper than the mylohyoid muscles are the geniohyoid and geniohyoglossus muscles attached to the genial tubercles on the inner side of the mandible If the lymphatic nodes at this point are enlarged they can be felt

The hyoid bone is next and it can be readily felt in the median line If it is not easily discovered in the median line it can be felt by a finger and thumb placed on each side of the neck above the thyroid cartilage

Passing over the hyoid bone the finger sinks into the space between the hyoid bone and the top of the thyroid cartilage Next comes the thyroid cartilage or Adam s apple This structure can readily be seen in adult males and in thin persons of either sex Despite obesity it can always be felt The finger then sinks into the space between the thyroid cartilage above and the cricoid cartilage below

From the cricoid cartilage which is on a level with the sixth cervical vertebra down to the sternum only soft structures can be felt The sternum projects forward and the trachea inclines backward so that opposite the top of the sternum the trachea is about ¾ inch (2 cm) behind the sternum The distance between the top of the sternum and the cricoid cartilage in an adult male is about 1¾ inches (4 5 cm) The trachea may be brought closer to the anterior portion of the sternum by overextending the head

If palpation is limited strictly to the midline only the isthmus of the thyroid can be felt However it is well to remember that the thyroid gland sometimes enlarges during menstruation and always somewhat during pregnancy returning to the normal size during the interval

A tumor in close relations with the trachea and moving with it during the act of swallowing is an enlarged thyroid gland A solid hard enlargement is usually a simple goiter and the enlargement ordinarily is greater on one side than on the other Determination of the presence or absence of induration and of fluctuation pulsation and systolic thrill and murmur is routine during physical examination of the gland

If fluctuation is present the enlargement is in all probability a cyst either of the thyroglossal duct or of the thyroid gland If pulsation thrill and murmur exist, the *case is one of exophthalmic goiter If a diffuse enlargement with tenderness* is present an acute or subacute thyroiditis is the cause

Tumor or aneurysm in the root of the neck and behind the sternum may cause prominence of the thyroid

The thyroid gland in some conditions of disease undergoes *atrophy* Often in myxedema and cretinism the thyroid is impalpable

Abnormal situation of organs in the neck often indicates disease for instance displacement of the trachea to one side or the other may be due to chronic fibroid phthisis thoracic aneurysm or mediastinal tumor In pulmonary tuberculosis the trachea is drawn to the affected side by the shrinking lung Aneurysm of the innominate artery pushes it to the left

Tracheal tugging is considered a valuable sign in cases of suspected mediastinal tumors or aneurysms In aneurysms it depends on the fact that the arch of the aorta curves back over the left bronchus and if an aneurysmal dilatation of the arch is present each systolic distention of the vessel presses down on the bronchus thereby pulling the trachea downward

To determine the presence of tracheal tugging if the patient is ambulatory stand behind him and have him close his mouth and raise his chin The tips of the fore fingers are then placed firmly on either side of the cricoid pressing upward then if a downward tug is present it will be detected

MUSCLES OF THE NECK

The sternocleidomastoid muscle is the one muscle of the neck that lends itself for examination If the patient is lying down have him raise the head, or if sitting up turn the head as far as possible to the right and left, with or without resistance offered by applying the hands to the sides of the head, or bend it forward against pressure on the forehead If the muscle is not paralyzed it stands out prominently (Fig 5-3) nerve supply, the spinal accessory nerve (eleventh cranial nerve)

Torticollis or Wryneck Torticollis or wryneck a disorder in which the neck is so twisted that the face is turned toward the side opposite the contracted muscle

and looks somewhat upward usually is caused by some disorder of the sternocleidomastoid muscle The sternocleidomastoid is not always the only muscle involved for the trapezius and others often are affected likewise owing to an attempt of the other cervical rotators to sustain and relax the sternocleidomastoid and thus obtain physiologic rest for it When the trapezius alone is involved the head is drawn backward and toward the affected side

Torticollis is congenital or acquired The congenital form is caused by injury to the sternocleidomastoid muscle occurring at birth or to developmental defects of atlas occipital bone or cervical ribs A swelling or tumor may be present in the course of the muscle when the muscle has been injured at birth In the acquired form the distortion may be more or less permanent and may be due to caries or other disease of the spinal column In such cases it is evident that treatment is to be directed to the diseased spinal column rather than to the sternocleidomastoid muscle which is found to be relaxed

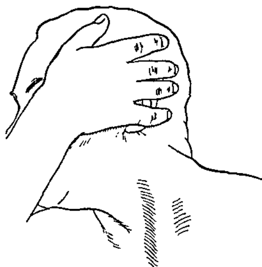


Fig 5 3 Sternocleidomastoid muscle At tempts are made to rotate the head against re sistance Nerve supply spinal accessory nerve (eleventh cranial nerve)

Inflammation of muscles fascia or the lymph nodes of the neck may cause the patient to hold the head and neck in a distorted position *wryneck* The wryneck in such cases will disappear as the cause subsides Rheumatic affections taking cold in the neck are a common cause of torticollis in which the sternocleidomastoid muscle may become contracted

A chronic torticollis occurring in an adult may be conversion hysteria

Spasmodic Disorders *Nodding spasm* a rhythmic nodding movement of the head and neck may be a form of habit spasm If nodding spasm is accompanied by a momentary loss of consciousness the condition is epilepsy Nodding spasm may be indicative of hysteria either alone or as a part of a rhythmic or hysterical chorea (salaam convulsion) Pulsating shaking of the head seemingly arising in the neck is often seen in the aged and occasionally in persons with aortic insufficiency

Spasmodic jerking of the head recurring every few minutes in which the head is brought toward one shoulder and at the same time the face is rotated to the opposite side and the chin raised is the clonic form of spasmodic torticollis The shoulder may be simultaneously jerked upward to meet the head In chorea the movements of the head are by contrast extremely irregular and bizarre and the facial muscles are affected before those of the neck

DISEASES DUE TO PRENATAL INFLUENCE

Tumors of the Thyroglossal Duct The vestigial rests of the thyroglossal duct account for some of the midline tumors of the neck These tumors lie between the isthmus of the thyroid gland and the base of the tongue Most commonly they occur either below or above the hyoid bone A remnant of the thyroglossal duct may end at the hyoid bone but should it continue upward it may pass in front of through or behind the body of that bone

Thyroglossal cysts are independent of the thyroid gland A cyst of the thyroglossal duct is manifested by a gradually developing painless enlargement in the

front of the neck If unsuccessful attempts have been made to remove the cyst most can be learned from the history prior to surgical operation Before there has been infection or surgical intervention, both the tumor and the skin over the cyst are movable

Cysts occur also just beneath within or on the dorsum of the tongue When they are large there may be disturbance of respiration as well as of deglutition These cysts may become malignant

Thyroglossal cysts may become infected and may suppurate from time to time apparently healing completely to discharge again in a few years without having been operated on From such repetitions of suppuration sinuses may result

The exact diagnosis of cyst of the thyroglossal duct can be made only by microscopic study of the tissues

Branchiogenous Cysts and Fistulas *Branchiogenous cysts* commonly occur in the upper part of the anterior triangle of the neck just beneath the angle of the jaw They develop slowly In the beginning these tumors are benign but they may become malignant while yet of small size In the early stage a branchiogenous cyst is not apparent unless the skin is stretched over it A tumor of this type can attain a large size displace the sternocleidomastoid muscle backward and bulge in the floor of the mouth at the base of the tongue

Branchiogenous fistulas are persistent gill cleft remnants remaining as ducts which extend from near the floor of the mouth to the skin in the region of the insertion of the sternocleidomastoid muscle around to the midline The whole of the duct or only part of it may be persistent If one of these fistulas is not patent at one end or at both ends it is a cyst and is called a gill cleft cyst

There is a history that the tumor opened spontaneously or by incision and that since its opening there has been a draining sinus There is a small inflamed tumor medial to the sternocleidomastoid muscle If a sinus is present it may drain continuously or it may become closed for some time to reopen anew

When one of these fistulous openings is explored with a probe it can be followed for varying distances in the direction of the floor of the mouth The probe may even pass the entire length into the oral cavity

The diagnosis is made by microscopic study of the tissue

Dermoid Cysts Dermoid cysts are small midline tumors of the neck which appear under the chin The patient usually complains of a bulge under the chin like the chin of an obese individual These tumors often bulge into the floor of the mouth or they may appear almost entirely within the mouth on each side of the tongue Other situations in the neck for dermoid cysts are near the mastoid process or the cysts may protrude from the mediastinum in the episternal notch

Diagnosis is established by microscopic study of the tissues

Angiomas There are congenital hemangiomas and lymphangiomas of the neck

Hemangiomas occur rarely in the cervical regions They usually are situated in the supraclavicular fossae They may be deeply set in the tissues and may not be discovered until years have elapsed These tumors often do not increase in size but remain as they were at birth The hemangiomas communicate with the venous system and consequently they are compressible as soon as the pressure is removed they reappear Some of these tumors are large and produce extensive bulging over the lower part of the neck Generally however they are limited to the skin or to the superficial tissues

Diagnosis is established by the behavior of the tumor on application of pressure The tumor disappears on pressure and returns immediately on release of the pressure

Lymphangiomas are common lesions of the neck which often are situated beneath the lower jaw and gradually attain a large size filling the entire side of the neck They occur also in the supraclavicular fossae and in the axillary and the

femoral regions Occasionally a lymphangioma may extend into the mediastinum As a result of irradiation these tumors may appear to be diffusely inflamed and under these circumstances are difficult to differentiate from neoplastic disease

The diagnosis is established by microscopic study of the tumor tissues

LYMPHADENITIS OF THE NECK

One of the commonest affections of the neck is inflammation of the lymph nodes The term lymphadenitis formerly was employed in descriptions of tuberculosis and syphilis The term as used here includes all infections of lymph nodes

The acute infections of the lymph nodes are caused by one or more of the pyogenic microorganisms *Spirocheta Borrelia Treponema* or a filtrable virus In the great majority of cases these acute infections of the lymph nodes subside and are designated as nonsuppurative lymphadenitis A nonsuppurative lymphadenitis is a regular concomitant of tonsillitis and other acute bacterial infections of the mouth Enlarged lymph nodes from viral infections do not suppurate

In infections of the mouth the lymph nodes in the drainage area of the mouth become enlarged and sensitive to pressure The reaction of the lymph nodes is usually obscured by the reactions of the causative disease The inflamed nodes may themselves produce general symptoms after the original causative lesions have subsided However the manifestations of an acute lymphadenitis usually are limited to tenderness due to the inflammation of the capsule of the lymph node and to perinodal edema If the infection is severe the capsule of the node may be invaded and if a number of nodes are involved the inflammation may extend beyond the capsule so that the individual nodes cannot be made out

The situation of the involved nodes depends on whether the infection is situated in the mouth or in the throat The enlargement of the nodes may be progressive along the line of drainage that is first those near the site of infection enlarge and become tender then they subside to be followed by enlarged nodes in more remote regions The involvement of the lymph nodes in remote regions of the neck or diffuse involvement is usually indicative that infection is not in the mouth Infection of the scalp the fingers arms and occasionally of the axilla may cause an enlargement of the lymph nodes in the neck At times it may seem that the acute inflammation of these cervical lymph nodes is going to terminate in suppuration the edema and cellular induration having formed a diffuse mass but almost always the lesions subside spontaneously

Acute Diffuse Lymphadenitis (Ludwig s Angina) Extension of the inflammation in an infected lymph node beyond the node or its capsule constitutes diffuse lymphadenitis Occasionally diffuse lymphadenitis will develop in the neck as a result of some severe infection in the oral cavity and this acute diffuse infection tends to follow the planes of the fascia in the neck This condition if severe is known as Ludwig s angina Ludwig s angina usually is confined to the floor of the mouth and because the tissues become very hard and woody the condition is known as woody phlegmon or ligneous phlegmon which later extends to the neck At times Ludwig s angina may be extremely acute and may terminate fatally within a very short period 1 or 2 days

Viral Lymphadenitis Many of the viral infections and infectious mononucleosis are accompanied by preaural and cervical enlargement of lymph nodes

Chronic Lymphadenitis The lymph nodes subsequent to an acute or an unrecognized inflammation may become chronically enlarged and be palpable as discrete nodules

In the presence of chronic lymphadenitis it is important to determine whether tuberculous infection exists and to differentiate it from other types of chronic lymphadenitis The beginning of tuberculous lymphadenitis is usually in the superior

triangles of the neck There is no acute onset The nodes become enlarged slowly

Tuberculous Lymphadenitis Tuberculous lymphadenitis is commoner in children than at other ages but it occurs frequently enough in young adults In an occasional instance of tuberculous lymphadenitis the lymph nodes break down and a sinus results Even in these cases recovery tends to occur spontaneously and often completely

In some instances in those in a weakened condition, tuberculous lymphadenitis develops rather rapidly The development is slow in those who have good health

In the beginning of tuberculous lymphadenitis the nodes are discrete but they tend to become matted together When these nodes remain discrete and are situated in the lateral triangles of the neck there is a resemblance to the lymphoblastomas especially of the Hodgkin type

Often a lesion is not suspected to be tuberculous until after it has been removed, cross section and microscopic study of the tissue then confirm the diagnosis

Lymph nodes situated over the parotid gland are particularly likely to break down and to leave sluggish superficial ulcers Once the skin is perforated a chronic discharging sinus ensues

Syphilitic and Mycotic Lymphadenitis In former years it was taught that syphilis was a common cause of lymphadenitis of the neck An enlargement of the lymph nodes in the posterior triangles of the neck if the possibility of pediculosis capitis had been excluded was considered to be almost pathognomonic of syphilis It is now known however that there are many causes of subacute chronic and indeterminate lymphadenopathies which include infections by bacteria viruses or fungi toxoplasmosis and diseases of unknown origin such as infectious mononucleosis

ULCERATIVE INFECTIONS OF THE NECK

Aside from ulcerating tuberculous lesions the common pyogenic boil or furuncle and fistulous tracts from congenital cysts there are very few ulcerating lesions of the neck Other than the lesions just named are those of the various fungi Blastomycotic ulcers on the neck usually are accompanied by ulcers elsewhere on the body especially on the hands Sporotrichosis likewise which is common on the hands may occur on the neck Actinomycotic infection of the mouth or jaws may occasionally cause ulcerations of the neck Other uncommon causes of ulcers of the neck are Vincent s angina leukemic tumors and abscess from the cervical vertebrae and dental infections

CAROTID SINUS SYNDROME

(*Vagal Syncope—Thomas Lewis Nothnagel s Syndrome*)

The regulation of blood pressure is an autonomic proprioceptive reflex mechanism which acts mainly by the intermediation of the pressoreceptor innervation of arterial and venous vascular areas (Weiss and Baker)

The bifurcation of the common carotid artery is richly supplied with sensory receptors which originate in the adventitia and leave the sinus as the sinus nerve to join the glossopharyngeal nerve Thus afferent direct nerve connection exists between the carotid sinus and the medullary centers

Weiss and Baker demonstrated that an abnormally sensitive carotid sinus mechanism can be responsible for attacks of unconsciousness and convulsions and that such attacks can be reproduced by slight trauma such as pressure over one carotid sinus The attacks are associated with cerebral anoxemia resulting either from cardiac asystole or from a primary reflex depression of the blood pressure A hypersensitive state of the carotid sinus reflex may produce unconsciousness convulsions or milder manifestations through one or more of three main reflex arcs The three motor pathways responsible for syncope involve the vagus nerve the vasomotor depressor nerves or the central motor pathways In a mixed form one or the other pathway may play the dominant role In normal

subjects mechanical stimulation of the carotid sinus does not produce symptoms The hyperactive carotid sinus reflex may be either unilateral involving only one sinus the activity of the other reflex remaining normal or bilateral

Three types of carotid sinus syndrome have been described In the *vagal type* the symptoms particularly the dizziness fainting and weakness result from cardiac asystole The asystole is due either to sino auricular or to auriculoventricular block which in its turn produces acute cerebral anoxemia In its pure form the afferent reflex travels over the vagus nerve to establish a heart block The *depressor type* usually appears in association with one or the other two types The symptoms result from primary reflex vasodilatation and secondary depression of the blood pressure entirely unrelated to cardiac slowing or any form of cardiac arrhythmia In the *cerebral type* the symptoms result apparently from impulses which travel directly to the brain The attacks may be induced within as short a period as 5 seconds Neither atropine nor epinephrine aborts or relieves the cerebral type of attack

The loss of consciousness as a complaint may occur under diverse circumstances such as nervous upsets claustrophobia change in position quick turning of the head tight collar or blows on the neck The reflex is more prevalent in men than in women The frequency and degree of response increases as age advances Coronary disease is the commonest condition in which the reflex occurs with the greatest frequency and the highest degrees of response

The loss of consciousness may come on suddenly or gradually There usually are pallor sweating severe weakness nausea and sometimes dizziness with a sensation of falling The heart rate may drop often to about 40 and the blood pressure falls rapidly The radial pulse is not palpable The unconsciousness is of short duration the patient is confused for a few minutes and weak for some time afterward Sometimes following loss of consciousness a convulsive seizure ensues as in Adams Stokes syndrome

If a hypersensitive carotid sinus is suspected an attack can be produced Before inducing an attack have the patient sit in a safe place to prevent injury in case he should fall The attack is then precipitated by pressure or firmly stroking the area of the carotid sinus

Vagovasal reflexes may bring about the same changes in some patients as those which follow swallowing with an inflamed or irritated pharynx or esophagus The induced attacks are not dangerous but are unpleasant and if frequent cause nervous worry which probably favors more attacks However an occasional death has been recorded after artificial stimulation of the carotid sinus

These patients should have electrocardiographic studies to rule out a heart block before attempts are made to induce an attack or to stop an attack by the use of drugs The attack should not be induced in any patient suspected of having an organic heart disease

PROTRUSION OF CERVICAL INTERVERTEBRAL DISKS

The sex distribution shows that affected males decidedly predominate A history of trauma may occasionally be obtained Before the appearance of symptoms of a medullary lesion signs of cervical radiculopathy may be present

The symptoms are extremely variable and resemble those of tumors and degenerative disease of the spinal cord Segmental symptoms arising from root compression may be present

The findings on physical examination are like the symptoms variable In advanced protrusions of a disk the findings are those due to pressure on the spinal cord in this region Plain roentgenograms of the cervical portion of the spinal column may show extensive spondylosis deformans Myelograms made by the injection of air into the cisterna magna reveal a subarachnoid space which is narrowed and distorted by an anterior protrusion which reaches the spinal cord There

is no obstruction to the passage of the air into the thoracic subarachnoid space In only a few cases will the protrusion be limited to one intervertebral disk

TUMORS OF THE NECK

Tumors of the Carotid Bodies These tumors are rare lesions which arise in the carotid bodies which lie in the angle formed by the branching of the common carotid artery into its two branches This angle is situated in the superior carotid triangle at a midpoint equidistant from the mastoid process the angle of the jaw and the thyroid cartilage Carotid tumors may develop however at considerable distances from this general situation They may be bilateral and familial These tumors grow very slowly their development parallels that of the mixed tumors of the salivary glands Pain transmitted to the region of the ear is occasionally present In most of the rare instances of carotid tumor the growth is discovered when the neck is being palpated for lymph nodes These tumors may become malignant the chances are four in five however that they will remain benign

Diagnosis is established by the presence of chromaffin cells at biopsy

Ganglioneuromas In Recklinghausen s disease (generalized neurofibromatosis) there are multiple tumors along the peripheral nerves all over the body Some of these tumors may be situated in the neck These multiple tumors usually found in the deep midportion of the neck grow very slowly (see Diseases of the Peripheral Nerves Tumors Chapter 6)

Diagnosis is suggested by the presence of Recklinghausen s disease but can be established definitely by biopsy

Primary Cystadenomas Cystic tumors of the neck comprise cystadenomas which arise in the lymph nodes and cystic aberrant thyroid glands The other cystic tumors which have been enumerated in the foregoing paragraphs are regarded as remnants of the gill clefts and cysts of the thyroglossal duct

Lipomas and Myxomas Lipomas of the neck are rare They do occur in the lower and posterior regions The myxoid tumors of the neck are more deeply seated generally than lipomas However good differentiation on palpation cannot be made between myxoid and lipomatous tumors

Diagnosis is established by biopsy study

Atheromatous Cysts In common with dermoid cysts atheromatous cysts or wens occur on the lateral surfaces of the neck especially on the posterior portion These tumors are definitely attached to the skin but their walls are distinct from the surrounding tissue

Lymphoblastomas *Lymphoblastomas of the Hodgkin type* involving the cervical lymph nodes are particularly difficult to differentiate clinically from tuberculosis A common location of palpable lymph nodes in Hodgkin s disease is along and behind the midportion of the sternocleidomastoid muscle and in the supraclavicular fossa These lymph nodes are often a conglomerate irregular mass rather diffuse tumor like and matted together In Hodgkin s disease each lymph node maintains its independence but the sense of touch may not interpret this individuality of existence of lymph nodes in a large mass which fills the whole region of the neck If Hodgkin s disease is suspected clinically often there are palpable nodes in the axillae and the inguinal regions because Hodgkin s disease or other diseases of the lymphoblastic group may arise first in these regions The beginnings of Hodgkin s disease may be in the mediastinum or in the retroperitoneal lymph nodes and the disease may be limited to these regions throughout its course with very few nodes enlarged in the neck axillary regions or inguinal region Hodgkin s disease may invade the skin

Diagnosis of Hodgkin s disease is established by biopsy of a node

The *lymphoblastomas of the sarcomatous type* are commoner in middle aged men whereas the lymphoblastomas of the Hodgkin type are commoner in young

men Lymphosarcomas appear in the upper parts of the posterior triangles of the neck or just beneath the upper third of the sternocleidomastoid muscle These tumors may begin in the anterior triangle of the neck and often in the supraclavicular fossa A common site of lymphosarcomas is in the mediastinum where they are discovered by means of roentgenograms usually after the tumors have invaded other tissues and some symptom is presented which requires roentgenologic examination of the thorax Lymphosarcomas often invade the skin or the tissues surrounding them and they grow rapidly

Diagnosis is established by biopsy of an enlarged node

Lympho Epitheliomas (Endotheliomas) of the Lymph Nodes A lympho epithelioma *usually appears at the angle of the jaw and often precedes or follows* a pharyngeal tumor a tumor of the cranial cavity or a sinus A characteristic feature of these tumors is that severe pain may attend the appearance of the initial enlarged node or even antedate its appearance

Lympho-epitheliomas are fixed in position from the beginning These tumors are highly malignant and the termination is an invasion of the neck and the mediastinum by direct extension

Diagnosis is established by biopsy of a lymph node

Malignant or Secondary Tumors Secondary tumors of the neck are metastatic tumors which occur within the lymph drainage system from a primary malignant tumor and correspond in structure to the original tumor It is not always possible to determine the site of the original tumor

For the most part however the secondary malignant tumors of the neck are carcinomas that have originated on the lips tongue gums floor of the mouth nasopharynx face or scalp In carcinoma of the stomach testicles or kidneys there may be a small fixed lymph node—*Virchow s node* or sentinel or signal node—superior to the medial end of the left clavicle

Melano-epitheliomas may occur on the lip or in the neck they metastasize to all parts of the body Metastatic tumors in the regions of the neck are prone to become secondarily infected from the oral cavity irrespective of the site of origin

DISEASES OF THE PHARYNX

The mouth the larynx the esophagus the two eustachian tubes and the two posterior nares open into the pharynx which thus serves as the common tract for intake of air food and drink

Pharyngeal Openings The opening of the *mouth* into the pharynx sometimes is narrowed from cicatricial contractions resulting from ulcerative processes such as syphilis and from burns due to swallowing of caustics There rarely is obstruction downward so that patients with such narrowing usually can swallow but the cicatrices contract the opening upward and the soft palate its arches and the walls of the pharynx may be bound together in one cicatricial mass preventing respiration through the nose

The opening between the *larynx* and the pharynx is easily accessible to those experienced in the procedure of examination If the tongue is drawn well forward the tip of the epiglottis can be seen and if a long straight tongue depressor is used in many cases the arytenoid cartilages and even a portion of the vocal cords can be seen

The opening into the larynx can be felt by a finger introduced into the mouth In cases of suffocation from a foreign body the object often is lodged at this point and thus part of the foreign body is in the larynx and part in the pharynx A foreign body can often be dislodged if the finger is thrust its full length into the mouth and throat and swept from side to side

The opening between the *esophagus* and the pharynx is in a line with the long axis of the pharynx The opening of the larynx on the contrary *is more on the anterior wall* of the pharynx It is for this reason that a tube introduced either through the mouth or through the nose passes into the esophagus and usually does not enter the larynx Excep

tions to this rule occur more frequently in comatose patients in whom the tube passes more freely into the larynx

The *eustachian tube* passes from the anterior portion of the tympanic cavity downward forward and inward to the upper posterior portion of the pharynx The eustachian orifices one on each side are on a level with the floor of the nose The upper border of the pharyngeal opening is approximately ½ inch (1 27 cm) above the soft palate and about an equal distance below the basilar process The lateral third of the eustachian tube near the ear is bony and the inner two thirds are cartilaginous The isthmus of the tube is at the point of junction of the bony and cartilaginous portions and is the narrowest portion of the tube The tube opens *in swallowing and yawning and admits air to the tympanic cavity and mastoid cells Interference with this process may cause the* so called aviation otitis with retraction of the ear drum during aeroplane trips Catarrhal affections of the throat readily travel up the tube and set up an inflammation of the middle ear Swelling of the lining of the tube follows and air no longer passes to the ear

The eustachian tubes are often *involved in acute or chronic infections of the throat* and middle ears The commonest symptoms related to eustachitis is popping and roaring noises in the ears These noises may be enhanced by or synchronous with the pulse beats

The *posterior nares* open into the upper portion of the pharynx above the soft palate This portion the nasopharynx differs from the oral and the laryngeal parts of the pharynx in that its cavity always remains patent

Pharyngeal Diverticula *In* pharyngeal diverticula the pouch opens into the pharynx by a transverse opening in the middle line about 1 inch (2 5 cm) in length The orifice varies in size in different cases and may be merely a slit The pouch makes its passage between the pars obliqua and the pars fundiformis of the inferior constrictor immediately above the mouth of the esophagus There are two hypotheses as to the origin of these diverticula the first that they are due to embryologic defects the second that the cause is mechanical *The theory that there is weakness in this region that is due to some sort of prenatal influence concerned in the formation of either type of pouch probably is correct*

The symptom that is always constant in a pharyngeal pouch is the return of fragments of undigested food This return is not immediately after the food has been taken but many hours or even days afterward Solids are more troublesome to swallow than liquids there is no dysphagia but the patient has to make two efforts before deglutition is accomplished Abnormal secretion of saliva and difficulty in disposing of it *may be the only sign in elderly persons There is no vomiting or pain but the patient may notice a gurgling noise in the throat especially when lying down and at meals this noise is followed by the bringing up of quantities of phlegm*

The diagnosis depends on the presence of fullness in a posterior triangle of the neck on which pressure produces a gurgling *noise* and the escape of gas into the mouth The opening into and the diverticulum itself *may be demonstrated by* roentgenologic and endoscopic examinations

Pharyngitis The mucous membrane of the nasopharynx is ciliated columnar that of the lower portion of the pharynx is squamous The mucous membrane contains racemose mucous glands and follicles or crypts surrounded by lymphoid tissue and it is well supplied with blood vessels The pharynx is frequently affected by inflammation or pharyngitis An abnormal general redness of or discrete eruptions on the palate fauces and pharynx may be due to simple inflammations of the mucous membrane measles scarlatina rubella (rotheln) roseola epidemic influenza erysipelas or irritant poisons The vesicles of varicella variola or herpes may also be seen So also with petechiae infarcts and extravasations of blood or bleeding surfaces

When the lymph follicles are markedly involved they can be seen studded over the posterior wall of the pharynx This constitutes follicular pharyngitis Not infrequently some ulceration may be present forming ulcerative pharyngitis Should abscess formation occur around the pharynx arising from an infection from the oral cavity the pus occupies the retropharyngeal space between the buccopharyn

geal fascia and prevertebral fascia Its spread upward is limited by the skull laterally by the sheath of the carotid vessels hence it passes downward behind the esophagus and may enter the posterior mediastinum

The Pharyngeal Tonsil (Adenoids) The pharyngeal tonsil composed mainly of lymphoid tissue extends across the posterior wall and roof of the pharynx be tween the eustachian tubes

Hypertrophy The pharyngeal tonsil when enlarged hangs from the vault of the pharynx It is a somewhat lobulated mass and when large in children obstructs nasal respiration Under these conditions the disease known as adenoids is present. Mouth breathing results and the child is likely to snore and make other queer respiratory sounds when sleeping These children may literally fight for breath at night

A child who has reached 3 or more years of age and has marked adenoids pre sents a typical appearance The mouth is open the lips are thick the upper lip is elevated the teeth project the chin recedes the alae nasi are atrophied and tense from want of use and the nasal orifices are reduced to elongated slits The philtrum is indistinct and disuse of the muscles between the upper lip and the eyes gives the face a flat smooth characterless expression Most such children have a more or less constant discharge of mucopus from the anterior nares leading to excoriation and eczema of the upper lip The nose however in some cases may be dry The adenoidal child often breathes noisily owing to obstruction The voice is often without resonance The most important secondary affection of adenoids is the effect on the organs of hearing According to some observers almost one third of these children have some defect in hearing The facial expression may remain after the adenoids have been removed and persist into adult life

Retropharyngeal Abscess Retropharyngeal abscess may arise from any one of three causes cervical caries suppuration of lymphatic nodes or extension of pus from the middle ear through the canal for the tensor tympani muscle Retropharyn geal abscesses occur external to the pharyngeal aponeurosis and bulge into the throat Because of the looseness of this aponeurosis and its lack of firm attachments these abscesses may not bulge forward as a distinct circumscribed swelling They are more likely to gravitate downward and hang in a loose baglike manner opposite the base of the tongue They are too soft to be easily felt They are more easily discovered by inspection In the search for the etiology of retropharyngeal abscess roentgenologic examination of the spinal column is made to detect the possible existence of spinal caries or Pott s disease and the ear is examined for suppurative otitis media

Diseases of the Pharynx Due to Disturbances of Innervation or Psychic Control *Anesthesia* of the pharynx is associated with hysteria and is indicative of some interference with the sensory functions of the glossopharyngeal and vagus nerves It is found in diphtheritic neuritis hysteria and bulbar paralysis Globus hystericus is a functional disorder of the ninth nerve

Spasm of the pharyngeal muscles on attempting to swallow is a functional affec tion of the motor portions of the same nerves It may be present in neurotic and hysterical individuals in hydrophobia true or false in tetanus and in strychnine poisoning It is probably an element in globus hystericus

Paralysis of the pharynx if bilateral causes difficulty in swallowing the food is not passed into the esophagus and portions may enter the larynx If the paralysis is unilateral there is little if any difficulty in deglutition Pharyngeal paralysis indi cates an involvement of the nuclei of the ninth and tenth cranial nerves or of the nerves in their course as in bulbar paralysis Landry s paralysis and neuritis or in basal lesions as meningitis tumor or aneurysm

Malignant Growths Malignant disease is rare as a primary affection of the pharynx It is usually secondary to direct propagation in the last stage of cancer of

the larynx the base of the tongue, a tonsil or the esophagus Either carcinoma or sarcoma may occur the former is the commoner

In malignant disease in the region of the pharynx the patient at first complains of discomfort in the throat and interference with speaking swallowing and nasal respiration Sharp pains develop early and extend to the neck ears or angle of the jaw Pain of burning character in the ear is a common symptom

On examination there often is observed the presence of frothy mucus or saliva in one or both of the pyriform fossae With the increase of the growth the voice may be hoarse and respiration is observed to be seriously interfered with There are salivation dysphagia blood stained sputum foul breath and enlargement of the regional lymph nodes

The age of the patient the one sided affection spontaneous pain and progress of the disease suggest the diagnosis It is established by biopsy study of tissue re moved

DISEASES OF THE LARYNX

The larynx is composed of the large epiglottis the thyroid cartilage the cricoid car tilage and three pairs of small cartilages

In the infant the larynx is situated opposite the second third and fourth cervical vertebrae In the adult it has descended to the level of the fourth fifth and sixth cervical vertebrae

The uppermost part of the epiglottis is below the dorsum of the tongue Extending forward from the epiglottis to the base and the sides of the tongue are three glosso epiglottidean folds In the fossae formed by these folds foreign bodies such as capsules tablets and fish bones may become lodged Objects lodged here are easily seen by aid of the laryngoscopic mirror

The vocal cords lie immediately behind or just below the most prominent portion of the anterior edge of the thyroid cartilage which commonly is called the Adam s apple

Examination of the Larynx Examinations of the larynx and the lingual tonsil to be of value require skill and considerably more knowledge of the procedures than can be gained by casual interest and an occasional attempt to make such an ex amination

The laryngologist may report the presence of swelling ulceration or tumors on above or below the vocal cords In tuberculosis multiple ulcers may be seen Syphi litic or cancerous ulcers are usually described as single ulcers In the ordinary forms of laryngitis the vocal cords are said to be of a gray pink or reddish color and pinkish nodules may be present on the edges of the cords A failure of the cords to move properly or paralysis may be reported

Laryngeal Paralysis Paralysis of the muscles of the larynx is rarely if ever due to cortical disease but is caused by lesions affecting the nuclei or origin of the vagus in the medulla or the nerve itself at some part of its course Thus it may be implicated at its origin by the lesions of bulbar paralysis syringomyelia locomotor ataxia or multiple sclerosis or an injury tumor or disease of the base of the brain may involve the nerve near its superficial origin The causes of paralysis most fre quently encountered are those which affect the vagus nerve or its laryngeal branches in the neck or the thorax namely in the neck enlarged cervical lymph nodes en larged thyroid gland tumors of the neck or the pharynx and caries of the cervical vertebrae in the thorax aneurysm of the aorta abscess tumor or enlarged lymph nodes in the mediastinum esophageal cancer and rarely compression resulting from inflammation of the pericardium or pleura and pulmonary tuberculosis The nerve and its branches may become inflamed (neuritis) or the laryngeal muscles themselves may undergo pathologic changes terminating in paralysis as an effect of the toxins of a number of the acute specific infections notably diphtheria or chronic poisoning especially from lead and alcohol

Total bilateral paralysis is most commonly indicative of aneurysm of the thoracic aorta cancer of the esophagus great enlargement of the thyroid or mediastinal tumor

Abductor paralysis is commonly unilateral and is the most frequent variety of laryngeal palsy

Adductor paralysis either bilateral or unilateral is as a rule of functional origin

Paralysis of the *interarytenoid muscle* (adductor) is most likely to be a symptom of hysteria

Paralysis of the *internal thyro arytenoid muscles* (adductors) is a not uncommon result of overuse of the voice and laryngeal inflammation

The Voice (The Faculty of Speech) A loss of voice or whispering voice (aphonia) and coarse or harsh quality of the voice (hoarseness or dysphonia) are both due to some interference with the function of the vocal cords Abolishment of action of the cords may be due to local and inflammatory disease of the larynx or to disease of or pressure on the recurrent laryngeal nerve Aphonia may come suddenly or be preceded by hoarseness or the latter alone may be present

In the majority of cases aphonia and hoarseness are due to some form of laryngitis such as the acute or chronic catarrhal variety associated with acute infections of the upper part of the respiratory tract syphilis or tuberculosis Edema of the glottis and retropharyngeal abscess are occasional causes Excessive use of the voice or excessive cigarette smoking may induce congestion of the cords and cause hoarseness or aphonia Tumor of the larynx impacted foreign bodies and cicatricial stenosis of the larynx may be found responsible

Disorders of Speech from Involvement of the Laryngeal Nerves The larynx receives its nerve supply from the superior and inferior laryngeal branches of the vagus nerve Interference with their functions gives rise to anesthesia of the larynx or paralysis of the laryngeal muscles manifested by aphonia hoarseness and if the abductors are affected dyspnea

If there is a deep hoarse voice and a brassy cough with a tendency for particles of food to enter the larynx there is probably interference with the superior laryngeal nerve giving rise to anesthesia of the upper portion of the larynx and paralysis of the cricothyroid muscles This interference is often due to bulbar paralysis a peripheral neuritis or the pressure of a tumor such as a goiter or carotid aneurysm

If there is aphonia without cough or dyspnea it may be due to paralysis of all the laryngeal muscles caused by bulbar paralysis tumors of the medulla diphtheria or pressure on both recurrent nerves or to complete paralysis of the adductors which is usually of functional origin but may occur from overuse of the voice or laryngitis

Hoarseness with easy fatigue on slight use of the voice is indicative of unilateral abductor paralysis of which the commonest cause is a mediastinal tumor or an aneurysm pressing on one recurrent nerve If right sided it may be due to a much thickened pleura cancer of the upper part of the esophagus or mediastinal tumor

A modified but understandable voice and phonation may be maintained in the presence of a dangerous laryngeal paralysis resulting from locomotor ataxia bulbar palsy or pressure from a tumor or aneurysm

A rattling rough voice the so-called ventricular voice is due to the occasional vicarious vibration of the false vocal cords or ventricular bands in cases in which the true cords are paralyzed or destroyed by ulceration Other abnormalities of the voice consist in a double or triple splitting of the sounds In the first variety the two sounds although simultaneous differ in pitch (diplophonia diphthongia) this phenomenon is due to a small tumor on the edge of one vocal cord or to unilateral paralysis of the cords The triple voice is very uncommonly observed and is caused by a pedunculated tumor which has its attachment below the vocal cords During

the early portion of phonatory expiration the growth rises, the voice is clear, durin the middle part it lies between the cords the voice is of normal tone but of decreased volume and during the latter part the tumor has been protruded to a point above the glottis and the voice is clear

The peculiar quality of voice which goes by the name of nasal voice is some times a matter of habit and lack of training of the voice It is of service to recognize *two varieties of the nasal voice* The first the *open nasal* is caused by nonclosure of the nasopharyngeal opening by the soft palate and may be imitated by speaking without opening the mouth the second the *closed* or *stopped nasal* is due to nasal stenosis and resembles the tone imparted to the voice by speaking while the nose is pinched between the thumb and forefinger

The open nasal tone is indicative of a congenital cleft of the palate of paralysis of the soft palate or destruction of the soft palate The closed nasal voice is often present in coryza hypertrophic rhinitis nasal polyp or postnasal adenoids

Dumbness or *mutism* inability or unwillingness to speak may be present in hysteria idiocy melancholia and dementia Feigned inability to speak is sometimes resorted to by malingerers A real inability to speak may be due to cerebral fatigue which succeeds a severe and exhausting illness particularly severe fevers Mutism exists in those who are congenitally deaf or in children who become totally deaf before the power of speech is permanently acquired

Anarthria indistinct or imperfect speech the impairment varying in extent may be an evidence of paralysis of the tongue soft palate and facial muscles Glossitis parotitis and the absence of teeth may be responsible for a mumbling manner of talking

A *slow interrupted* manner of speech the words being slurred over somewhat as if the patient were intoxicated with tremulousness of the tongue and lips is observed in general paresis and myxedema

A *piping querulous* voice with manifest hesitation in beginning a sentence the words then being rapidly spoken is characteristic of paralysis agitans

Scanning speech in which the words are spoken slowly each syllable accented as if reading verse is an important symptom of multiple sclerosis but is also found in Friedreich s (hereditary) ataxia

These disturbances of the voice may be of such nature that their final diagnosis and differentiation require a consideration of the faculty of language

The Faculty of Language Normal Speech Mechanism The normal exercise of the faculty of language depends on the existence and integrity of certain cerebral centers (*1*) *psychic centers of intelligent perception* (*2*) *sensory receptive centers* (*3*) *emissive or motor centers* and (*4*) *the association tracts* Finally there are the basal ganglia for the special senses and the nuclei in the medulla and spinal cord actuating the muscles employed in articulate speech which constitute respectively the sensory and motor peripheral apparatus

Apraxia If an individual has varying degrees of defects in the power of recog nizing or understanding the nature and uses of objects or possibly the identity of persons the condition is termed apraxia The varieties of apraxia correspond in number and character to the varieties of sensations mind blindness mind deafness mind anosmia mind ageusia and mind atactilia

Apraxia is closely allied to and is often found in connection with, the various forms of aphasia that is an inability to produce or understand spoken or written speech (see The Nervous System and Locomotion Chapter 6)

Aphasia and Its Varieties The term aphasia embraces a variety of defects in the use or in the comprehension of language either spoken or written In order to understand and interpret this symptom and its several forms a conception of the normal manner and mechanism of the faculty of language is desirable The subject is complex and many exceptional and often unexplainable cases occur As far as

possible the statements made here are those the truth of which is considered as probable

MOTOR OR ATAXIC APHASIA This variety embraces as its most important division *aphemia* the loss of power to utter words although the patient knows what he desires to say because he cannot revive the motor memories for articulating them Occasionally these patients may be able to sing even though they fail in attempts to talk This sort of aphasia is common to most aging persons—those 50 years or more of age—in a mild form *Agraphia* is inability to write words for lack of power to recall the motor memories of the muscular actions involved in writing

SENSORY APHASIA The principal varieties of sensory aphasia are the auditory and visual *Auditory aphasia* or word deafness is as its name indicates an inability to recognize words when spoken The patient hears but does not understand as if listening to a foreign language *Visual aphasia* or *alexia* is a condition in which the patient sees but does not recognize or understand written or printed words *Memory aphasia* consists of inability to recall the sound of a word or phrase Once the sound is recalled the word or phrase can be spoken This is a form of sensory aphasia incident to aging

EXAMINATION FOR APRAXIA AND APHASIA To determine the presence of *word deafness* (auditory aphasia) the examiner must elicit the information indicated by the following questions

1 Can sounds be heard by the patient? Ability or lack of ability to hear determines the presence or absence of ordinary deafness

2 Can spoken words be heard by the patient? If so then

3 Can the spoken words be understood? This may be ascertained by having the patient put out the tongue open and close the hand A failure to comply indicates inability to attach any meaning to the words and word deafness is present which is the principal symptom of auditory aphasia

To determine the presence of *word blindness* (alexia visual aphasia)

4 Can the patient see objects?

5 Can written words be read?

6 Can written or printed words be understood? This may be ascertained by writing out a question or a direction for instance Close your eyes A failure to comply indicates word blindness which is the principal symptom of visual aphasia

To determine the presence of *motor aphasia* (aphemia)

7 Can the patient speak voluntarily?

8 Can words be repeated?

9 Can the patient read aloud? If unable to do these things motor aphasia or aphemia is present

To determine the presence of *agraphia*

10 Can the patient write?

11 Can dictation be written?

12 Can a sentence or words be copied? If he is unable to do these things agraphia a symptom which may be found in all forms of aphasia is present If the patient is unable to write voluntarily because of inability to remember words or their appearance but can write to dictation the disability is sensory agraphia If it be impossible to write either voluntarily or to dictation a motor agraphia is present

To determine the presence of *mind blindness*

13 Are ordinary objects recognized and their uses known? Put before the patient two or three familiar objects and question him about one of them If the proper article is not picked up the subject has mind blindness provided that inability to understand the request is owing to word deafness

To determine the presence of *mind deafness*

14 Does the patient recognize ordinary sounds that is does the ring of a bell the ticking of a watch awaken a recognition of its import? If not the patient has mind deafness

There are variations in the degree of all the speech defects There may be only a

the early portion of phonatory expiration the *growth rises, the voice is clear, during* the middle part it lies between the cords the voice is of normal tone but of decreased volume and during the latter part the tumor has been protruded to a point above the glottis and the voice is clear

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tracheitis acute obstructive laryngotracheobronchitis and an unclassified noninfectious croup due to allergy foreign body and chemicals or other irritative agents) The presence or absence of supraglottic edema and the position and motion of the vocal cords before and after suctioning of the subglottic area and trachea have furnished a dependable guide to the choice of therapy

Here the interest is in spasmodic croup

Spasmodic croup is a form of laryngeal stridor of unknown etiology characterized by sudden onset and unaccompanied by fever It affects children more commonly boys than girls between the fourth month and the end of the second year of life The attacks may continue until the child is approximately 10 years of age

The disease affects feeble undernourished children who often have nasopharyngeal adenoids enlarged faucial tonsils enlarged bronchial lymph nodes and disorders of the upper air passages

The child as a rule goes to sleep as usual He suddenly is awakened from a sound sleep struggling for breath The forced effort at inspiration produces a croupy crowing stridor In the effort to fill the lungs the child sits up in bed tosses his hands about holding to the nearest support or clutching his throat All the accessory muscles of respiration are thrown into action The mouth is open the nostrils are dilated and the chest is heaving but as all these efforts fail to fill the lungs the diminished pressure causes retraction of the lower ribs and the manubrium sterni The expression is one of terror as the patient becomes cyanotic The head is thrown back and the child is soon covered with sweat In severe attacks respiration may cease for a few seconds In such cases there are carpopedal contractions convulsions and even involuntary passage of urine and feces The attacks are usually of short duration often less than a minute and rarely do they exceed 2 minutes The attack is ended by one long deep relieving inspiration Relief from the attack is followed by a croupy cough Further attacks may take place during the same night and they often recur from night to night for some time After the attack the child falls asleep and the next morning his health is as usual

The diagnosis of this condition is not difficult if it is remembered that the paroxysms characteristically begin with sudden onset and without fever and hoarseness and end as abruptly as they begin

Syphilis Ulcers of syphilitic origin may involve almost any portion of the larynx but they usually are anterior and involve the epiglottis These ulcers often are associated with syphilitic manifestations in the mouth which are ulcers that are most marked posteriorly blanching of the mucous membrane of the mouth and the presence of a white frothy mucus The serologic reactions for syphilis are positive

Stenosis Acute laryngeal stenosis is met with in wounds injuries foreign bodies acute laryngitis septic inflammation diphtheria membranous laryngitis the exanthems and syphilis It has been known to occur rather suddenly in tuberculosis of the larynx

The chief symptoms of laryngeal stenosis are dyspnea stridor and alteration or extinction of the voice All of these symptoms are intensified on exertion The patient prefers to sit up in bed for breathing seems easier in this posture It is remarkable how hard a patient may be striving for air in this condition and yet be free from marked cyanosis

The diagnosis is established by laryngoscopic examination

Laryngeal Symptoms of Nervous Origin The laryngeal symptoms in mediastinal tumor are due to irritation of or pressure on the vagus nerve or its recurrent laryngeal branches There are cough attacks of dyspnea and alterations in the voice In the irritative stages the cough is paroxysmal hoarse imperfect and brassy When one or both recurrent branches are completely paralyzed the cough loses its sharp ringing character and becomes hollow and blowing

partial loss of any one of the normal powers of expression depending mainly on the extent of the productive lesion

DISEASES WHICH CAUSE APHASIA In the majority an aphasia is one of the symptoms of organic focal cerebral disease occurring in the left hemisphere in right handed persons and vice versa The localization and appraisal of aphasia is the work of the neurologist Aphasia may occur as the result of a cerebral hemorrhage thrombosis embolism abscess or tumor

In rare instances aphasia occurs in hysteria and neurasthenia and epilepsy It is an occasional symptom in migraine and may be present during convalescence from severe and exhausting fevers

Infections of the Larynx ***Acute Laryngitis*** Acute laryngitis is an acute catarrhal inflammation of the mucous membranes of the larynx manifested by hoarseness or aphonia and occasionally by cough The acute inflammation may primarily attack the larynx whence it spreads downward to the trachea and bronchi or upward to the nose Frequently the laryngitis is but an extension downward of an acute catarrh of the nose or the pharynx Or rarely an acute inflammation first develops in the bronchi and spreads upward to the larynx The attack often is precipitated by excessive and, more particularly improper use of the voice It may follow on the vomiting and retching of an alcoholic debauch an attack of seasickness or a passionate attack of crying or sobbing

In the early stages of acute laryngitis the visible changes in the larynx may be very slight but as the hoarseness increases the signs of inflammation increase proportionately

When uncomplicated an acute laryngitis is without danger and subsides within from 3 to 10 days

Chronic Laryngitis Chronic laryngitis is a catarrhal inflammation of the mucous membranes of the larynx *characterized by an impairment and alteration* of the voice Disorders of the nasal and the postnasal cavities and to a less extent of the pharynx and mouth are the most potent factors in originating chronic laryngitis The alteration of the voice is at first an intermittent huskiness but when the symptom is well established the hoarseness is persistent it is more marked after a rest or on arising in the morning than at other times and tends to disappear after a little use of the voice The tone of the voice is lowered aphonia is seldom complete except after prolonged or extreme forcing of the damaged organ A sense of fatigue and soreness of the throat may be present There is frequent clearing of the throat by hemming and hawking

On physical examination the mucous membrane is congested The vocal cords may be only slightly affected but often appear dull and gray

The diagnosis usually is not difficult for the examiner who is trained in inspecting the larynx Because tuberculosis of the lung may be accompanied by a chronic and inveterate laryngitis care should be taken to exclude the possibility of this disease

Nodular laryngitis is a form of chronic laryngitis generally caused by faulty use of the voice and characterized by local thickening of the vocal cords The symptoms are those of chronic laryngitis The diagnosis is made by one who is accustomed to inspect the vocal cords and larynx

The laryngologist may describe other forms of laryngitis such as (1) hypertrophic laryngitis and (2) chronic atrophic laryngitis

Laryngismus Stridulus or Spasmodic Croup Gilbert and associates have called attention to the multiplicity of names for croup and offer the following classifications (1) diphtheritic croup with three subgroups (diphtheritic obstructive laryngitis laryngotracheitis and laryngotracheobronchitis) and (2) nondiphtheritic croup with five subgroups (acute catarrhal laryngotracheitis supraglottic edematous obstructive laryngitis subglottic obstructive exudative and edematous laryngitis and

CARCINOMA OF THE VOCAL CORDS Four of each five laryngeal cancers arise on the vocal cords Cancer of the vocal cords is almost always either squamous cell carcinoma or papillary carcinoma The low inherent malignancy of these neoplasms in the presence of poor lymphatic drainage of the true cords renders carcinoma arising on the cords likely to remain localized for a considerable period of months occasionally longer

The cardinal and only early symptom of carcinoma of the vocal cords is hoarse ness Hoarseness in middle aged or aging men which persists for more than 2 weeks requires inspection of the larynx

A general physical examination does not reveal any significant findings so long as the laryngeal tumor is limited to the structure Early evidence of extension is recorded by the otolaryngologist as immobility Only in extensive spread will there be any palpatory evidence present

All lesions of the larynx are examined for evidence of syphilis

Examination of the thorax may aid in differential diagnosis in regard to tubercu losis and syphilis Roentgenograms of the larynx may be helpful in indicating the site of origin position and extent of laryngeal tumors

Biopsy is necessary to establish the identity of any proliferative or ulcerative lesion of the cords Benign papillomas may cause confusion even on microscopic examination Repeated biopsies are occasionally necessary There is no evidence to indicate that carefully performed biopsy on the vocal cords increases the danger of spread of cancer

Surgical treatment is the preferred procedure when the lesion is limited other wise radium may be used

Leukoplakia Laryngis Leukoplakia of the larynx is characterized by white thickenings of the upper surface of either or both of the vocal cords that appear much the same as the patches of leukoplakia of the tongue Leukoplakia laryngis can be diagnosed only by a physician of a great deal of experience in examinations of the larynx

DISEASES OF THE TRACHEA

Acute Tracheitis Acute tracheitis is usually a part of the common cold al though in some instances an acute infection seems to be limited to the trachea

The symptoms of acute tracheitis vary in intensity and the duration is com parable to that of acute bronchitis The symptoms consist of hoarseness cough and perhaps fever and ineptitude The diagnosis is by inference if the patient promptly recovers the voice and if the cough and the fever cease

Chronic Tracheitis A chronic tracheitis often of severe grade frequently arises from a long-continued inhalation of irritating fumes or dusts or from exces sive tobacco smoking Chronic infection of the paranasal sinuses mouth nose and throat when the discharges are aspirated into the trachea frequently results in chronic tracheitis and bronchitis

An endoscopic examination may be necessary as diagnostic proof if the rest of the respiratory system is normal

Tracheal Stenosis Irritating gases corrosive fumes and acute and intense infections may cause stenosis of the trachea which may reach such a degree as to result in severe dyspnea and extreme cyanosis This often occurs during the course of an ordinary bad cold

Tumorous growths inside or pressure from outside the trachea may cause a chronic slowly progressive stenosis Trauma and slowly progressive infections such as that of syphilis also may produce stenosis

Rupture of the trachea usually due to the trauma of instrumentation as a rule is fatal When tracheal rupture occurs from trauma of other origin the condition

In tabes dorsalis there may be laryngeal anesthesia paresthesia or hyperesthesia The symptoms are described respectively as tickling irritation, and tightness or a sensation of choking The voice becomes thick and jerky owing to ataxic movements of the cord Laryngeal crisis often occurs in the early stage and may precede other signs of the disease by several years The laryngeal crisis and spasmodic attacks vary much in intensity and frequency they seldom cause death by asphyxia

Other diseases of the nervous system which may affect the larynx are disseminated sclerosis bulbar palsy dystrophia myotonica (myotonia atrophica) myasthenia gravis and syringomyelia

The diagnosis is suspected when there are attacks of dyspnea alterations in the voice and cough during the course of the aforementioned diseases

Laryngeal Vertigo Laryngeal vertigo (laryngeal syncope laryngeal epilepsy) occurs in middle aged neurotic men who are suffering from laryngitis bronchitis bronchial asthma or pulmonary tuberculosis The attack begins with laryngeal tickling or irritation followed by a short cough spasm of the larynx dyspnea transitory syncope and slight convulsive movements Such attacks may recur every day or at intervals of a month or more

Laryngoscopic examination is impossible during the attack and the findings are negative in the interim The diagnosis is made from the history and the presence of an obvious psychoneurosis Serious deterioration of health may follow the onset of laryngeal vertigo

Tumors Benign Tumors Benign tumors of the larynx are comparatively rare In this group the *papilloma* is the commonest It occurs earlier than any other type of benign tumor and may be congenital Nearly all laryngeal growths met with in infants and in children up to the age of 10 years are of this character

CYSTOMA Pathologically cysts of the larynx are due to retention from obstruction in the duct of a muciparous gland Their commonest situation is on the anterior surface of the epiglottis

OTHER BENIGN TUMORS Lipoma is a rare tumor of the larynx as are angioma adenoma myxoma lymphoma neurofibroma and tumor of the thyroid gland The last named tumor which may arise from abnormal distribution of thyroid tissue is extremely rare and of relatively little importance

LARYNGEAL POUCH This pouch is conveniently referred to here because it clinically resembles a benign tumor although its pathologic origin is uncertain Laryngeal pouch the so called prolapse of the ventricle of Morgagni is a membranous sac between the superior vocal cord and the thyroid cartilage On examination a smooth pink fleshy growth with a broad base is seen issuing from the ventricle of Morgagni and resting on the vocal cords

Malignant Tumors Both carcinomas and sarcomas occur in the larynx In appearance and symptoms carcinoma and sarcoma of the larynx are so much alike that biopsy must be performed to establish the diagnosis

Carcinoma of the larynx may occur without involvement of the vocal cords or these structures may be primarily affected

Lesions of the larynx which do not involve the vocal cords have their origin in the epiglottis the false cords the ventricular cavity or the subglottic area These lesions cause virtually no early symptoms and are usually not discovered until there is pain, extensive infiltration of cartilage and muscle or metastasis to the regional lymph nodes With the occasional exception of lesions localized on the tip of the epiglottis pain is seldom a complaint until the disease is well advanced and then pain often is severe extending to the ear and the side of the head and being much increased by swallowing Respiratory obstruction is marked when these intrinsic lesions advance There may be pharyngeal symptoms and interference with deglutition These lesions may not be amenable to resection but many are radiosensitive

During the active secondary stage of syphilis changes in the trachea and bronchi may occur resulting in symptoms of an acute tracheitis or bronchitis Mucous patches develop and in the tertiary stage gumma is sometimes seen in the trachea and bronchi Cough is usually the first symptom of this condition The gumma may be absorbed so that the cough does not become productive If ulceration occurs however considerable mucopurulent sputum is produced From such ulcers there often is blood streaked sputum or even frank hemorrhage These ulcers occasionally extend through the wall of the trachea or a bronchus and invade the adjacent structures such as the mediastinum the esophagus and even the pulmonary artery Stenosis of various degrees results in difficulty in respiration even to the point of terminating fatally In such cases the physical findings including results of roentgenologic examination do not differ from those caused by stenosis from other causes

Diagnosis is sometimes assisted by the detection of syphilitic disease or its scars in the nose pharynx or larynx but in many cases these aids are not forthcoming and then the cause of the stenosis is arrived at by inspection or by endoscopy

The prognosis will depend on the degree and the depth of the stenosis In cases in which the contraction is low down or cannot be relieved or extends into the bronchi the outlook is very grave

Syphilis of the Lungs O Leary was able to prove the diagnosis of syphilis of the lungs in only 6 instances over a period of 20 years

Syphilis of the lungs is so rare that the symptoms are unknown

The diagnosis is made by exclusion There are nodular infiltrated lesions in the upper pulmonary fields positive results of serologic tests of the blood and negative results from other diagnostic laboratory procedures A therapeutic test for syphilis is said to confirm a presumptive diagnosis when the symptoms disappear roentgenologic examination shows that the lesions in the lungs have disappeared and the patient gains weight and comments on how well he feels The serologic reactions of the blood do not show any significant change until many years later

Tumors Benign Tumors The benign growths of the trachea are papilloma fibroma lipoma adenoma hemangioma and lymphoma lymphadenoma mixed parotid tumors osteoma enchondroma and aberrant thyroid tissue tumors Papillomas and fibromas are as in the larynx the commonest Papillomas of the trachea the most frequent growths of all are generally extensions from the larynx and hence occur in young children Fibromas come next in frequency

If the growth is in the trachea symptoms may be noticed only when there is considerable stenosis The patient usually first complains of dyspnea This may occur suddenly and fatally The voice is usually enfeebled though not lost Hoarseness will depend on the degree to which the vocal cords are involved by catarrh Simple neoplasms of the bronchi generally attract attention in the first place by causing hemoptysis Dyspnea is not severe but obstruction to the escape of secretions from the lung produces cough and expectoration of infected sputum sometimes of fetid character

The effects are mechanical owing to lack of aeration and drainage of the lung beyond the point of obstruction The neoplasm in such case may occlude the trachea It may however if pedunculated be pulled down during inspiration and cause obstructive emphysema by its reversed check valve action

If there are manifestations of obstruction of the trachea information may be gained by roentgenography

Amyloid Tumors of the Larynx and Trachea The larynx and the trachea are common sites of localized deposition of amyloid Amyloid tumors of the larynx trachea or bronchi may occur in association with amyloid degeneration elsewhere in the body or as isolated deposits The isolated deposits according to New may be divided into (1) diffuse subepithelial infiltrations (2) tumor forming amyloid deposits and (3) amyloid degeneration in a pre-existing tumor

usually is not suspected because of the other serious symptoms resulting from the trauma Spontaneous rupture rarely occurs Tracheo esophageal fistula is fortunately rare These fistulas cause severe respiratory distress and dysphagia

In summary all of these conditions namely acute tracheitis chronic tracheitis tracheal obstruction and stenosis and tracheo esophageal fistula may be diagnosed clinically However roentgenoscopic and bronchoscopic confirmations are obtained when warranted by the conditions

Acute Laryngotracheobronchitis Acute laryngotracheobronchitis occurs in children and in the aged *Streptococcus hemolyticus* and *Hemophilus influenzae* are causative organisms There is a sudden onset of cough dyspnea and cyanosis A tenacious and thick later purulent exudate is coughed up The tenacious secretions may obstruct bronchial ramifications and thus produce atelectasis Mediastinal emphysema and spontaneous pneumothorax may result from overdistention of the alveoli On examination of the chest there are noisy coarse rales but no changes on percussion or in the breath sounds

The diagnosis is established by the history and the physical findings Roentgeno scopic examination in either children or the aged differentiates this condition from bronchopneumonia

Ulcerative Tracheobronchitis The tracheobronchial ulcerations may result from recurring attacks of atypical pneumonia bronchiectasis abscesses tubercu losis sarcoidosis and fungous infections The characteristic symptom is persistent cough with considerable blood streaked sputum which occasionally contains gross blood Fever and loss of weight or failure to gain weight are often present Exami nation by physical means usually gives negative results

The diagnosis is established by seeing the ulcerations with the bronchoscope The etiologic diagnosis may be made from materials obtained for pathologic and bacteriologic studies The prognosis depends on the etiology

Syphilis of the Trachea Bronchi and Lungs Congenital Syphilis This dis ease may occur in the larynx trachea bronchi and lungs of the fetus and the new born child Since the tracheal lesions may be the commoner and all of these struc tures may be synchronously affected they are all considered at this time

Acquired Syphilis Syphilis attacks any part of the larynx the vocal cords the trachea and the bronchi Mucous patches may appear in the larynx Tertiary laryn geal lesions are rare However they have been reported to result in partial or com plete stenosis of the larynx Deep ulcers sometimes occur and the disease may extend so as to result in external fistulas

SYMPTOMS Often there are hoarseness and difficult breathing when edema be comes severe Loss of voice may occur in severe infections Laryngeal scarring some times results in permanent difficulty in breathing

DIAGNOSIS When the disease is diagnosed by the history positive serologic reac tions the appearance of the lesions and biopsy and if the patient is treated promptly before much destruction has occurred the prognosis is good There may remain unfortunate conditions such as stenosis of the larynx

Syphilis of the Trachea and Bronchi Erythema mucous patches gummas perichondritis and subsequent cicatricial stenoses occur in the trachea and bronchi Diffuse gummatous infiltrations are commoner than the localized gumma They are most frequent at the lower end of the trachea near its bifurcation next in frequency come extensions from the larynx and lastly those situated midway between these points Syphilitic ulceration and stenosis may extend into the main bronchi Compli cations occur from perforation of the esophagus aorta pulmonary artery or vena cava but the commonest sequela is stenosis

Syphilitic ulcerations of the trachea are considered to be of rare occurrence Syphilitic stenosis of the main bronchi without a similar condition in the trachea is extremely rare

In the larynx amyloid deposition is almost entirely associated with a generalized amyloidosis Though laryngeal symptoms occur these usually can be detected and identified as a part of the generalized lardaceous process The lesion occurs more frequently in men than in women in about the ratio of 3 to 1

The majority of patients are between the ages of 50 and 70 years at the time the diagnosis of amyloid tumor is made In cases reported in the literature the youngest patient was 19 years of age and the oldest patient was 80 years old

SYMPTOMS When symptoms are present they are caused by the physical presence size and location of the tumor There are hoarseness and dyspnea Willmann reported a case in which the diagnosis was made 6 weeks after the onset of symptoms Thompson described a case in which the chief symptom of hoarseness in a woman 32 years old had been present at least 25 years before the diagnosis of deposition of amyloid was made The usual duration of symptoms before diagnosis has been between 1 year and 2 years

EXAMINATION The gross appearance of the amyloid tumor has been said to be a waxy translucent yellow or yellow gray swelling without ulceration of the overlying mucosa The lesion has been reported to have either a smooth or a nodular outline and to be either diffuse or well localized and red The tumor is hard on palpation

DIAGNOSIS The diagnosis of an amyloid tumor is determined by microscopic examination of sections of tissue from the lesion The extent of the lesion may be determined visually by laryngeal examination roentgenologic examination tracheoscopy or bronchoscopy Evaluation of the extent of a recurrent lesion or a lesion of which the diagnosis has been established may be simplified by staining of the amyloid by the intravenous injection of Congo red

Malignant Tumors Malignant disease of the trachea is not common Most of the malignant growths present in the trachea are but extensions from the larynx esophagus or mediastinum They are much commoner toward the upper end of the trachea than toward the lower end and more often affect the posterior wall than the anterior wall

The early effects of malignant neoplasms are similar to those of innocent growths in the air passages namely dyspnea cough and expectoration with hemoptysis

In regard to frequency of hemoptysis in nontubercular disease of the trachea and lungs Jackson and Diamond found the principal causes to occur in the following order of enumeration bronchiectasis primary bronchial carcinoma tracheobronchitis pulmonary abscess no evident disease nonsuppurative pneumonitis suppurative pneumonitis bronchial adenomas secondary pulmonary cancer primary tracheal carcinoma The manifestations of a pulmonary abscess may be produced with severe toxemia An extensive growth produces cachexia pain if spreading into the mediastinum and dysphagia if involving the esophagus

The physical and roentgenologic examination will reveal the situation of the tumor in most cases but it is generally necessary to make a bronchoscopic examination which also affords an opportunity for biopsy

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The thin ones of postures 3 and 4 (Fig 6 1) suffer with the functional disturbances known to affect the visceroptotic type Those who have postures like those of 3 and 4 (Fig 6 2) are hypersensitive about both their posture and their degree of obesity

In many families it seems that posture *is heritable*

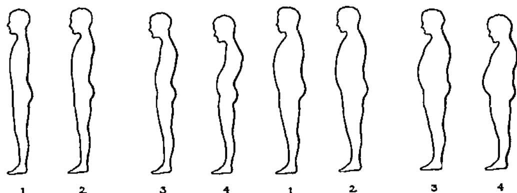


Fig 6 1 Postures of persons of normal or less than normal body weight Posture 1 is excellent 2 fair 3 poor and 4 bad

Fig 6 2 Postures of the obese Posture 1 is excellent 2 fair 3 poor and 4 bad

Dynamic Posture Dynamic posture is posture in motion or in action or in preparation for action It includes the transitions between the static positions of lying sitting and standing and also such activities as pushing lifting walking running climbing jumping dancing swimming work and play Dynamic posture includes the uses of the upper extremities and the trunk as well as of the legs and the relationships between the various parts of the body in action Dynamic posture includes locomotion which requires the combined use of all four extremities Poor dynamic postures are characterized by awkwardness and inability of performance and are present in diseases of the central nervous system

The basic dynamic posture is characterized by a slight crouch with the ankles knees and hips flexed the head and trunk inclined forward and the trunk slightly flexed the arms relaxed and slightly flexed With the body in this position the muscles are in a midposition with increased tone balanced and ready for instant and powerful action in any direction They act also as springs absorbing shocks and initiating movement

Walking Walking may serve as an example of dynamic posture The body is tilted slightly forward from the basic standing position and the weight is thrown on the ball of the foot while the other thigh is lifted and the leg and opposite arm are swung forward Various muscles aided by inertia maintain the body in balance on the ball of one foot until the opposite heel strikes when the weight quickly advances to this leg with the knee extended Momentum carries the body forward over the extended leg until it passes the perpendicular when the thrust of the foot renews the action and the process is repeated with the opposite foot and leg (Fig 6 3)

Correct walking is done with a smooth rhythm the muscles contracting gently with a brief wavelike action and relaxing in the interval It is characterized by free muscle and joint action momentum balance and rhythm Effort becomes much greater if the speed is increased or if momentum and rhythm are disturbed

Alertness nimbleness speed smoothness and steadiness are required for the development of dexterity Dexterity is the requisite for the synchronization of the movements necessary for the performance of proficient and deft walking especially at a military cadence of 120 steps per minute These attributes are obtained by (1) a heritage of good muscles and (2) a lifetime of careful training and control

6

DISEASES AFFECTING LOCOMOTION

Diseases affecting locomotion interfere with the modes of the body such as posture gait equilibrium and often positions while in bed at rest and during illnesses These modal disturbances result from and are the manifestations of diseases of the skeleton the peripheral vascular circulation muscles and nerves

POSTURE

The term posture refers to the sum total of the positions and movements of the body Static posture refers to the different positions of rest and the variations of these positions Dynamic posture refers to the body in motion or in action It is by means of these postures that the body attains comfort mechanical efficiency and physiologic function while at rest and during locomotion Disturbances in posture are frequent accompaniments of diseases affecting locomotion

Static Posture Static posture is the posture while at rest It includes the normal positions for lying sitting and standing *Lying* is more easily assumed than any other position Static posture may be affected by diseases of the skeletal structures and also by diseases of the heart and lungs

Sitting The usual sitting position is with the trunk and head erect and centered over the pelvis or tilted slightly forward with a medium or slight lumbar arch and with the hips and knees flexed at a right angle The sitting posture may be the only comfortable posture available to those who have congestive heart failure

Standing In the standing position the body is vertical and essentially straight when seen from the side as well as from the back The position is maintained with the spinal column rather than the shoulders The body achieves its full height in this position with the head and chin level not tilted back The weight is slightly more on the heels than on the toes although in the dynamic position of expected forward movement the weight shifts toward the toes A strong man or woman may faint if required to stand rigidly for a long time (30 minutes or more) because of the relatively static condition of the circulation and the accumulation of blood in the vessels of the lower extremities Ankylosis of both hip joints in extension limits the postures to those of standing and lying

The length of time that a person can stand without fatigue is related among other factors to the posture A person of good posture as represented by postures 1 or 2 (Fig 6 1) can stand much longer without fatigue than a person of poor posture Postures 3 and 4 (Fig 6 1) represent those of persons who have poor muscle tone from lack of exercise or from disease

Obesity changes the postural appearance Those who have good postures such as postures 1 and 2 (Fig 6 2) may not be handicapped while they are young However, as age advances postures 3 and 4 (Fig 6 2) are assumed

Postures 3 and 4 (Figs 6 1 and 6 2) will not permit proper space for good physiologic functioning of the organs of respiration and digestion Posture is always an important diagnostic consideration in diseases of the heart and lungs

During youth if the person is otherwise healthy poor posture may not be measurably detrimental to the state of health However as the years advance those of poor posture are noticeably more prone to poor health than persons of good posture

shoes corns rheumatism gout sciatica metatarsal neuralgia hip joint or knee joint disease or injury (recent or old) sacro iliac disease sprains inflammatory disease of the extremities short leg and paralysis of one leg give rise to a limping or hobbling gait The most characteristic methods of abnormal progression are seen in diseases of the nervous system

Ataxic Gait In walking the foot is raised suddenly and too high the leg is thrown forward with unnecessary vehemence and the foot is again brought to the ground heel first or flat footed with a stamp The feet are usually planted wide apart and while they are in the air they move as if the patient were doubtful where to put them The body is bent forward and the eyes are fixed on the ground in order to *supplement as far as possible the loss of muscular and articular sensation This gait* is extremely characteristic of locomotor ataxia

Cerebellar Ataxic Gait The manner of progression resembles that of an intoxicated person The patient walks with short steps and with his feet wide apart staggers reels sways to and fro and reaches a set point by zigzagging toward it The swaying is relieved if he is supported by the hands of the observer placed under the armpits This gait is significant of a tumor of the vermis or middle lobe of the cerebellum and is often called the titubating gait or simply cerebellar ataxia A somewhat similar gait is seen in Friedreich s disease hereditary cerebellar ataxia dementia paralytica ataxic paraplegia labyrinthine disease and to some extent vertigo from any cause

Steppage Gait This variety of gait is due to paralysis of the extensor muscles of the foot whereby when the foot is lifted its anterior part tends to hang or drop down In order to prevent the toes catching and tripping against the ground the leg carries the foot somewhat forcibly forward raising it at the same time unusually high thus throwing the toes upward and bringing the foot to the ground heel first Steppage gait resembles the gait of a man who is walking through snow thick grass or brushwood It is evidence of peripheral neuritis of the anterior tibial nerve and because of a certain resemblance to the gait of locomotor ataxia is sometimes termed the pseudotabetic gait

Spastic Gait The legs are rigid and move stiffly and there is apparent difficulty in bending the knees In consequence the foot is dragged along the toes catching and scraping on the ground In some instances owing to spasm of the adductors of the thigh the legs and knees touch and cannot be separated causing cross legged progression that is the legs and feet overlap at each step This gait depends on the excessive tension and spasticity of the muscles arising from lesions of the upper motor neurons It is therefore when bilateral significant of sclerosis affecting the lateral pyramidal columns of the cord The mode of walking in hemiplegia is a unilateral form of the same gait The paralyzed leg by a tilting of the pelvis is swung outward and around to the front the toes often scraping the ground

Festination The head and body are bent forward and the patient takes short shuffling hurried steps his speed tending to increase as he progresses exactly as if he were being constantly pushed forward and were trying to prevent it This gait is termed festination or propulsion In some instances if the patient is pulled rather suddenly backward he will take a number of backward steps (retropulsion) although the body remains in its forward leaning attitude This gait is characteristic of Parkinson s disease and extrapyramidal neuron disease

Waddling Gait The shoulders are thrown back the back is hollowed (lordosis) and the abdomen protuberant the body sometimes actually leaning backward In walking the feet are planted wide apart and the body swings from side to side at each step—the waddling or duck gait It is a very characteristic symptom of pseudohypertrophic muscular paralysis It is seen also in disease or congenital dislocation of both hip joints

over the motive parts involved in dynamic posture A lack of these attributes may indicate disease

Walking with a *limp* may result from pain weakness stiffness or *deformity* *Pain* may be caused by an injury such as a sprain fracture or bruise a blister, a corn or an inflammation such as arthritis Weakness may be due to many things, such as lack of sleep improper diet illness heat and humidity a poor heart lung disease or nerve or muscle damage Stiffness may be due to muscle or joint damage or to tense or contracted muscles Deformities such as knock knees bowlegs short or twisted

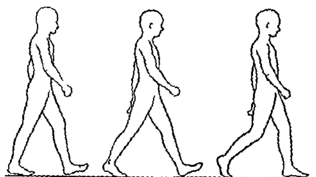


Fig 6 3 Walking Taking one step forward

legs clubfeet and high or relaxed arches have their individual effects on manner of walking

The security of balance and ease of walking or running are enhanced by swinging the opposite arm and leg forward and backward with each step as if in four footed locomotion The normal co ordination of movements of the extremities may be either exaggerated or lost when there is a limp

Poor Posture Poor posture may be due to changes in muscle tone or to changes in skeletal structures Irrespective of the cause a poor sitting posture is characterized by a drooping of the spinal column and trunk with the lumbar and thoracic portions of the spinal column flexed the pelvis tilted back the abdomen and chest flattened and compressed and the shoulders and head forward A poor standing posture is characterized by a forward movement and tilt of the pelvis, with an increased lumbar lordosis and an increased thoracic kyphosis In this poor standing posture the knees flex the abdomen sags and protrudes the chest flattens and the shoulders head and neck are advanced The height is diminished and the body as seen from the side forms a long S curve

Muscular Tone The tone of the muscles has a great deal to do with posture There is the large group of persons who have hypotonic muscles and elongated ligaments with joints which extend too far These hypotonic persons may have weak arches knock knees bowlegs torsion wryneck stooped backs curvatures of the spinal column and weak lumbosacral joints any one or a combination of which affects posture Any disease or injury which causes tonus changes in the muscles ligaments or bones is often the cause of poor posture In addition poor posture may result from poor training or poor habits

Gait The manner of walking is closely associated with the manner of standing In order to observe the mode of walking the examining physician requests the patient to walk away from and back to him to walk at right angles to the line of sight and to walk along a crack between the floor boards or a seam of the carpet

In pregnancy ascites large abdominal tumors cretinism pseudohypertrophic paralysis and obesity the body leans backward and the feet are widely separated so as to increase lateral balance while walking

Painful or disabling affections of one or both lower extremities such as tight

fevers such as typhoid fever even at an early period because of the marked muscular weakness and mental apathy so characteristic of typhoid fever It may be present in motor apathy which often attends frontal lobe brain tumors

RIGID DORSAL POSITION IN BED A rigid dorsal position with both legs drawn up in order to diminish abdominal tension is the rule in general peritonitis and in many cases of pelvic peritonitis In appendical peritonitis the right leg alone may be flexed In hip disease also there is flexion of one knee

ON SIDE IN BED The patient may lie on the side and the manner of lying may be active or passive as in the dorsal position There are many persons who cannot rest on one or the other side because of discomfort or actual pain Often no assignable reason can be found to account for this manifestation of discomfort

Patients having acute one sided affections of the thorax usually lie on the affected side to limit its movements and lessen the pain caused by pleural friction as well as to afford greater freedom of compensatory motion to the healthy side Moreover if a large pleural effusion exists the pressure due to its weight will not burden the heart and the healthy uncompressed lung when the patient lies on the affected side The posture of a patient with a cavity in the lung is of some significance that position being chosen which brings the opening of the cavity uppermost thus allowing secretions to accumulate and be discharged at infrequent intervals instead of constantly trickling into the bronchial tubes and causing an incessant irritating cough The rule of lateral decubitus is not invariable since the patient may find other positions preferable In sciatica the subject usually lies on the unaffected side

The lateral position with the legs drawn up to meet the trunk (the coiled posture) is seen in meningeal cerebral and cerebellar disease (due to spasm) and in hepatic renal and intestinal colic

OPISTHOTONOS Opisthotonos is an uncommon dorsal position in which the body rests on the head and heels the trunk being arched upward It is observed in strychnine poisoning tetanus and uremia as well as in some peculiar manifestations of hysteria A modification of this position is observed in the characteristic contraction of the posterior neck muscles in meningitis whereby the back of the head bores into the pillow

EMPROSTHOTONOS Emprosthotonos is an attitude in which the upcurved body rests face downward on the forehead and feet It is the opposite of opisthotonos and is rarely seen but may be observed in tetanus strychnine poisoning paralysis agitans and cerebrospinal meningitis

THE SKELETON

The diverse activities to which mankind is adaptable are first of all dependent on skeletal design The skeleton is composed of the axial and the appendicular divisions The axial skeleton is as the name suggests the center or the axis about which the movements of the appendicular skeleton are resolved

The *axial skeleton* embraces the bones of the head the spinal column and ribs the hyoid bone and the breast bone The spinal column includes the vertebrae of the cervical thoracic and lumbar regions and the sacrum and coccyx

The *appendicular skeleton* embraces the bones of the extremities including the shoulder girdle or thoracic girdle formed by the scapula and clavicle to which the upper extremities are attached and the pelvic girdle formed by the innominate bone to which the lower extremities are attached

In the four footed position the appendicular skeleton is concerned only in locomotion In the upright position assumed by man locomotion is more complex The upper extremity is freed from its main function of locomotion to follow the will of the brain In order to subserve the hand in the performance of its diverse functions the back a part of the axial skeleton has assumed more elasticity in order that it may bend to carry the hand to its appointed tasks

Equilibrium Equilibrium is a state of balance or equipoise a condition in which opposing forces exactly counteract each other Clinical histories of those who have sustained injury from sudden loss of lateral equilibrium are common Lateral equilibrium is often lost while one is carrying a weight in one hand *However injury to* the back may be sustained without a loss of lateral equilibrium if a heavy weight is carried in one hand and the pace is hurried or lengthened

Lateral equilibrium can be disturbed by a deviation of the spinal column above the sacrum and to one side and also by anything that affects the length of either leg When a position of rest is desired the hips are moved laterally so that the center of weight falls on one leg which is kept extended The opposite hip then descends until further adduction is stopped by the superior ligaments of that hip

In order to maintain balance the movements of the body must be properly performed and a relationship of the parts one to the other must be maintained This must be true whether the body is in a state of motion or of rest During motion the muscles control the position of the bones During rest the position of the bones is controlled by the ligaments The weight of the body acts as a constant force pressing downward and it is nicely balanced on the bones by the ligaments aided by the muscles A weakness of any one of these three elements ligaments muscles and bones may permit balance to be altered

If balance is altered, deformity ultimately will result Distortion of a bone as the result of a badly united fracture throws the weight and muscular action too much to one side with the result first that action of the part is impaired and second if use of the part is persisted in deformity results and increases If there are relaxation and weakness of ligaments during adolescence and if the ligaments give way in the foot for instance flatfoot results in the knees knock knee results and if in the back scoliosis or lateral curvature If the muscles give way as in poliomyelitis deformities result which may have to be corrected if they can be rectified at all by the proper application of braces or by tendon transplants

Mode of Moving In certain ailments there is unusual *immobility* due to the increase of pain on motion *as in arthritis scurvy and rachitis or a disinclination to* move because dyspnea is made worse by exertion Paralysis or tonic spasm of large muscular groups is another cause of enforced quietude

An opposite condition *restlessness* exists in many diseases as in fevers large hemorrhages and hyperthyroidism Agitated and irregular movements are seen in chorea in hysteria with its manifold manifestations and in other diseases of the nervous system Severe griping or colicky pain gallstone colic or renal colic will induce the patient to throw himself about in the hope of relieving his suffering

Posture in Bed Rest in Bed The patient usually takes to bed in acute illness or in chronic ailments because of general weakness pain or inability to remain up and around In some cases the patients may assume that they are unable to be up though they do not have pain weakness or fever In some special cases there is interference with the use of the limbs as in certain diseases of the nervous system

Many persons in health habitually assume certain attitudes while in bed and their customs in this respect may not be changed by illness *Because of this fact* the postures assumed in disease and their diagnostic associations are described as follows

Static Posture in Bed In the dorsal strong or active posture the patient lies on the back comfortably and without constraint This posture is seen *in health and* in slight illness unattended with great pain

Passive Posture in Bed In the dorsal inert or passive posture the patient lies on the back but is constantly slipping toward the foot of the bed *thereby putting the body in a posture* which is uncomfortable and which interferes with the respiratory movements The passive posture is observed in conditions of great weakness most frequently in the acute infectious diseases It is especially characteristic of severe

fibrous tissue The disks are attached to the cartilaginous plates of the vertebral bodies and are pulpified centrally to cushion the vertebral column for instance on jumping and landing suddenly on the feet

The skeletal muscles control the motion of the back within the limits determined by the ligaments These muscles are arranged in symmetrically opposing groups Posteriorly the short and long intervertebral muscles attached to posterior and lateral processes the vertebrocostal vertebro-iliac and vertebrosacral groups are extensors of the back and acting in unison with other groups rotation and lateral bending are accomplished

The tissues of the back are innervated and nourished by the posterior divisions of the segmental nerves and blood vessels

Variations and Anomalies of the Vertebrae Each human vertebra is formed by the fusion of adjacent halves of two primitive somites and passes through membranous and cartilaginous stages to become bone

Irregular fusion of the half somites gives rise to hemi vertebrae a frequent anomaly When it occurs there is usually a corresponding anomaly on the opposite side of the column a few segments distally which corrects the order of fusion Though the lesion causes deformity of the back it is of no great clinical importance

In the human vertebral column the number of segments of the vestigial coccyx and of that part of the sacrum distal to the articulations with the ilia is of no particular consequence The number of cervical segments is found to be remarkably constant The thoracic vertebrae distinguished by attachment of ribs vary in number from 11 to 13 the modal number being 12 The number of lumbar segments varies from 4 to 6 from a mode of 5 There is frequently a gain or loss of one thoracic segment with compensatory loss or gain of a lumbar segment the total number of pre sacral segments remaining unchanged Evolutionists aver that in man there has been a progressive shortening of the presacral column which has been synchronous with specialization in skull brain and teeth

The first sacral segment of the vertebral column is determined by the level at which the ilia articulate with the column to form the sacro iliac joints These are formed by the approach of the ilia developing in the limb buds to conjugate with the vertebrae The meeting is usually but not always bilaterally symmetric Lagging of one ilium behind the other results in asymmetric sacralization of the last lumbar or first sacral segment

The partial loss or gain of a segment as in the case of unilateral or bilateral sacralization of the last free segment with large transverse processes impinging on or articulating with the sacrum or ilium narrowing of the lumbosacral disk or formation of a false promontory rarely causes disability

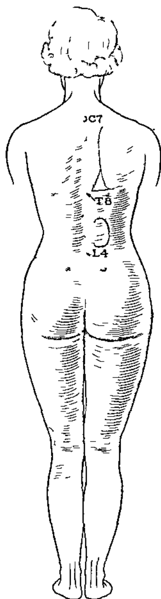


Fig 6 4 Topography of the back C7 seventh cervical vertebra *T8* eighth thoracic vertebra L4 fourth lumbar vertebra

Defects in closure of the neural canal and in development of the articular processes which anchor the lumbar column to the sacrum may occur In such instances the supporting structures of the sacrum may lie in any position between the sagittal plane characteristic of the lumbar vertebrae and the transverse plane of the sacrum and frequently these defects vary bilaterally and may definitely affect the stability of the lumbrosacral anchorage It often predisposes to a forward displacement of the involved vertebra on the segment below This forward displacement is known as spondylolisthesis

Skeletal Anomalies Anomalies of the axial skeleton comprise first of all those of the head These anomalies are great and mainly are incompatible with life Individuals who survive with head anomalies often have mental derangements (see Complaint Chapter 2 and Chapter 4) The rest of the anomalies of the axial skeleton are for the most part related to the number and development of the vertebrae and thus will be mentioned in relation to the vertebrae

Anomalies of the appendicular skeleton are related to the extremities The limbs either may fail to develop at all or may be present as mere rudiments of structure (amelus) Sometimes the proximal segments of an extremity are normal while the distal portion is deficient and tapers to a stump (hemimelus) In the reverse condition at least the proximal segment is missing and the hand or foot springs directly from the trunk (phocomelus) Union of the legs produces the mermaid condition (sympodia) A number of deformities may be associated when the hands or feet lack digits for example split or cleft anomaly of the hand and comparable deformities of the feet Opposite in nature is a partial or complete duplication (dichirus) An example of dichirus is *polydactylism* which usually is characterized by the addition of but a single digit The bony fusion or fleshy webbing of digits (*syndactylism*) is usually limited to union of the middle and third digits Abnormal shortness of the digits is *brachydactylia* due either to the omission of phalanges or to digital shortening The opposite tendency is hyperphalangism in which supernumerary phalanges are interpolated in the customary digital series All of these malformations of hands and feet tend to be strongly heritable (Arey)

Clubhand or *clubfoot* may result from primary defects in the differentiating limb buds with malformed muscles

Congenital elevation of the shoulder results from an arrested descent of the upper limb from its cervical embryonic position Congenital dislocation at the hip joint results from failure of the outgrowths that normally produce a brim about the socket floor of the joint *Intra uterine amputations* (at any level) are due to focal deterioration and not to constrictions by loops of the umbilical cord or amniotic bands (Arey)

THE AXIAL SKELETON AND LOCOMOTION

The Back and Spinal Column On inspection of the back of a normal person who is undressed and standing erect a median depression is observed to commence at the lower part of the line of the scalp and to extend almost straight down the back to the sacrum (Fig 6 4) The prominence of the seventh cervical vertebra and the first thoracic vertebra can be seen In the middle of the depression the spaced elevations representing the spinous processes of the vertebrae often can be seen and always can be palpated They form an approximately straight line These prominences end at the beginning of the sacrum

From the superior border of the sacrum down to near the top of the gluteal folds is a triangular space with its base above and apex downward The apex of this triangular space marks the third sacral vertebra Laterally opposite the second sacral vertebra the posterior superior iliac spine is palpable The elevations limiting the apex of the triangle are formed by the erector spinae muscles

Laterally from the furrow in the thoracic region the projections of the scapulae are visible If the arms are by the sides the borders of the scapulae are parallel to the median line The inferior angle of the scapula is opposite the upper border of the eighth rib The medial angle covers the second rib but its tip is level with the first rib The twelfth rib usually projects to just beyond the outer edge of the erector spinae muscle The outer edge of this muscle is marked by a depression separating it from the abdominal muscles in front

On the inner side of the lower third of the posterior edge of the scapula is a small triangle The upper side of this triangle is formed by the trapezius its lower side by the latissimus dorsi and its lateral side by the posterior edge of the scapula As the lung is nearest the surface at this point this triangle is often chosen for puncture

The spinal column is composed of a complex arrangement of bones joints muscles and fasciae Between each two of the presacral vertebral bodies is a disk of specialized

poliomyelitis It is often the result of the contraction of one lung after pleurisy or empyema

The development of scoliosis shifts the center of gravity from the midline to one side so that it falls nearer to the foot on the side toward which the trunk is inclined than to the contralateral foot (Fig 6 4) To compensate for this shift of weight the hips are inclined to the opposite side and the center of weight is brought by this inclination once more midway between the ankles However if the single curve of scoliosis is complete and again reaches the median line as is usually the case in scoliosis the center of weight may not be disturbed and if not lateral shifting of the pelvis does not occur There are instances in which the curves are so irregular that more of the weight shifts to one side than the other when this takes place the pelvis shifts and the hip on the side opposite to the inclination appears to be higher than the other (Fig 6 5)

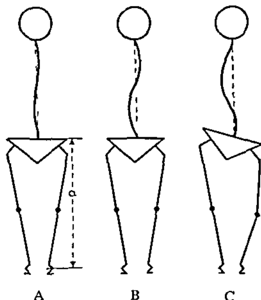


Fig 6 5 Deviations of the spinal column above the pelvis and measurements of the length of the legs

The line joining the two anterior superior iliac spines should be at a right angle with the long axis of the body as in A and B before measurements are made

If a hip is ankylosed its femur is moved laterally by the examiner until the line joining the anterior superior spines is at a right angle to the body The opposite femur is made to correspond in position before measurements are made

The line a (the distance from the anterior superior spine of the ilium to the tip of the internal malleolus of the tibia) when the foregoing conditions have been met gives the length of the leg in question This is compared with the same measurement of the opposite leg

Legs of unequal length cause the pelvis to tilt down on the side of the short leg as in C

Measurement of the Lower Extremities Under ordinary circumstances in the absence of deformity or limping the lower limbs are assumed to be of equal length When on measurement one can detect difference in length of the legs (1 inch or more) there usually is enough deformity to produce unevenness of the gait for instance a limp (Fig 6 5)

If the legs are of unequal length the pelvis tilts to the side of the shorter limb carrying the lower part of the spinal column with it and producing a convex curve again on the side of the short leg which is not marked in the lumbar region The middle and upper parts of the spinal column are carried to the opposite side of the convex curve In patients thus variously affected the deformities may be great In some the spinal curvature may extend as high as the shoulders and in these the hips may be observed to be uneven in height The unevenness in the length of the legs may be such that the unequal length is obvious and the gait is markedly limping

Backache and Pain in the Back Without Sciatic Radiation There is more good evidence to indicate that backache can be definitely associated with misuse and abuse of the back than there is that it results from an upright posture and plantigrade gait However it is recognized that the anatomic rearrangements required to change from a pronograde animal to one with an upright posture and a plantigrade gait were extensive From evolutionists (Sir Arthur Keith) we learn that these

More difficult to recognize is spina bifida occulta in which there is no external mass The patient *may have no complaints or may complain of slight motor* sensory or trophic disturbances in the lower extremities Over the lower part of the back in the middle line there may be an area of hypertrichosis On palpation a defect of the vertebral spinous processes in this region may be made out

At the lower end of the sacrum there is sometimes an abortive tail This may be a true tail *containing rudimentary vertebrae* sometimes the appendage does not contain bone

It is generally felt that complete sacralization of the last lumbar vertebra is not a cause of pain but that partial sacralization of this vertebra produces pain through irritative changes occurring at the site of the false joint These irritative changes may occur solely in the soft tissues or eventually they may cause osteoarthritic changes to take place at the site of pseudoarthrosis

Curvatures of the Spinal Column At birth there are a thoracic and a sacral curve of the spinal column Both of these are convex posteriorly Laterally there is a slight curve in the thoracic region with its convexity to the right or the left de*pending on the handedness of the individual*

If the height of the two shoulders in the adult is unequal the lower shoulder usually indicates the handedness usually the right shoulder is the lower in right handed individuals A marked difference in height of shoulders increases the chances that a rotary scoliosis is present

The symmetric or anteroposterior curvatures of the spinal column are kyphosis convexity backward and lordosis convexity forward An asymmetric or lateral curvature is scoliosis As a rule there are moderate kyphosis of the thoracic region and moderate lordosis of the lumbar region Practically the lateral curvatures or scolioses *are the most important of the abnormal curvatures*

Curvatures of the spinal column may result from faulty development or may arise from diseases of the spinal column itself Faults of skeletal development such as supernumerary ribs or absence of a rib may cause scoliosis Spondylitis is often a cause of kyphosis or of scoliosis Curvatures may develop in the course of Paget s disease acromegaly or syringomyelia Ordinarily curvatures of the spinal column depend on softening of the bones of the spinal column due to rickets early or late or to osteomalacia

Kyphosis especially in the thoracic region is seen in many elderly persons It forms part of the changes which constitute an emphysematous thorax A kyphotic condition may be present also in rachitic children and in young persons who are weakened by acute or long continued illness These posterior curvatures are to be distinguished from the more or less sharply angled kyphosis of Pott s disease In some patients children in particular there is a somewhat unusual but normal *prominence of the seventh cervical vertebra or the eighth and ninth* thoracic spinous processes

Lordosis occurs usually in the lumbar region and is an exaggeration of the normal curve in this region It may be due to the dragging weight of pregnancy ascites and all large abdominal tumors It is likely to occur in the course of muscular dystrophy In rare instances excessive lordosis of the lumbar region in adolescence is associated with orthostatic albuminuria

Scoliosis is a rotary lateral curvature of the spinal column with prominence of one scapula If one scapula projects it indicates the existence of lateral curvature *and the existing* causative conditions or paralysis of the serrator anterior muscle on that side If both scapulae are abnormally projecting it is probable that the patient has an alar or a pterygoid thorax

Scoliosis may indicate unequal length of the lower extremities the habit of standing habitually on one leg or carrying weights on one side muscular weakness from fever or anemia *or general debility congenital or acquired* It may also result from the *tilting* of the pelvis in old sciatica and from paralysis due to anterior

shows great limitation of back motion Spasm is present and percussion reveals localized pain and tenderness over the affected vertebra or vertebrae

Early in the course of the disease the diagnosis is difficult to make on the basis of the roentgenographic findings Later it is somewhat more easily made The shadow of paravertebral abscess may indicate an infectious lesion In general granulomatous lesions such as tuberculosis cause destruction of bone and cartilage and the bone has little or no tendency to heal until late in the course of the disease Active pyogenic infections cause early destruction of bone Proliferative bone healing occurs readily and there is little actual decrease in the width of the vertebra or interspace

The differential diagnosis of the infectious organism causing destruction is diffi cult when there is no history of a preceding disease Blood cultures may help in the early stages Complement fixation tests and agglutination tests may aid in those in whom the infection is due to *Salmonella typhosa Salmonella paratyphi* or *Brucella abortus* Tuberculin tests may be of negative value in diagnosis Occasionally aspira tion of the abscess and subsequent bacterial culture and animal inoculation will establish the etiologic diagnosis

Trauma In any injury to the spinal column or its structures which has not in jured nerves or the spinal cord the patient walks carefully without jarring This caution is greatly increased if the sacro iliac joints have been injured

In severe soreness of a sacro iliac joint the gait is suggestive The patient carefully avoids putting weight on the affected side of the pelvis either while walking or while sitting In the sitting position the weight of the body is maintained on the opposite ischial tuberosity

Strain Back strain indicates an applied stress greater than that which the sup porting elements are normally able to withstand If the tissues of the back are ab normal at the time of injury there may be an invasion by an inflammatory process and convalescence may be delayed Soreness stiffness and weakness persist longer than the seriousness of the injury would justify

In acute traumatic back strain there is localized tenderness with muscle spasm and pain The posture will be that of giving way to the injured side in order that tension may be relaxed on the affected side The pain may extend down the back of the hip and thigh or around to the inguinal region

In back strain the findings of roentgenographic examination are negative

Flat Back Strain Flat back strain occurs in the pyknic type of body (Fig 6 2 posture 4) The strain develops in the lumbosacral region with lordosis absent in the sitting standing and supine positions and with all activities limited by pain which is relieved only when the patient lies supine with the legs outstretched Stoop ing forward in the sitting position may be free but recovery in the upright position is labored and hyperextension is impossible because of pain The side lying and the prone positions are uncomfortable The roentgenogram reveals an unusually vertical sacrum

Sprain The term sprain is applied to a joint that has been suddenly twisted or wrenched thereby stretching the ligaments until they become torn or separated from their attachments Severe sprains are much rarer than are strains

After a back sprain there is immediate disability and the symptoms are more severe than in case of strain There may be severe suffering accompanied with nausea and faintness In severe sprain there is muscle rigidity and there is a defi nitely localized region of deep sensitiveness and tenderness There may be noticeable swelling over the region of the damaged tissues

A sprain of the back may be expected to heal as quickly as sprained ligaments of other joints but a residual soreness due to fibrous hyperplasia and often adhesions or scar tissue may prevent work for several weeks after the primary healing has

changes have imparted structural weaknesses These structural weaknesses which seem to have commenced in the caudal end of the spinal column involve most of the muscles and the corresponding nerve centers which control them In contrast to the weaknesses is the formation of the sacrum which serves to shorten and strengthen the spinal column

The whole weight of the suprasacral part of the human body is supported erect on the spinal column over long periods of time The demands on the neuromuscular postural mechanism are even greater in the sitting position than in the standing posture particularly if the individual sits leaning forward as in writing or reading The muscles which act on the short levers of the spinal column yield first while the muscles which act on the long costal levers can still keep on The human being rests these exhausted short spinal muscles by allowing the vertebrae to rotate until the articular processes begin to lock and the transverse processes rest on the neck of the long costal lever Therefore it is more restful at least for short periods of time to sit somewhat slumped forward It is an erroneous idea that sitting practically upright is less exhausting

Backache in the lumbar region or lumbago is a common discomfort in those who are acutely or chronically ill and in those who have overworked Backache may be due to causes within the confines of the spinal column or it may be due to causes situated within the supporting structures of the spinal column And finally it may result from disease of the viscera in the thoracic abdominal or pelvic regions

Backache is not a disease only a symptom that may be due to any one of different causes

Postural Backache The position assumed in poor posture of advanced degrees is familiar to all The head protrudes the shoulders are rounded and dropped the chest is flat and the thoracic portion of the spinal column has an increased posterior convexity or kyphosis The lumbar lordosis is exaggerated and the abdomen is protuberant particularly in its lower half The pelvic inclination is increased Some persons have a flattened lumbar lordosis and compensation takes place by flexion of the hips The knees are slightly flexed and the feet are pronated All degrees of these postural aberrations are encountered

Many people habitually assume at work or at other times positions of great mechanical instability and sooner or later under the stress and strain backache results

One common cause of chronic strain in the lower portion of the back is a sagging or protuberant abdomen (Fig 6 2) which by its weight and its downward and forward pull tires the muscles and leads to increased tension on the ligaments supporting the lumbar portion of the spinal column The postural backache of pregnancy is explained on the same basis

The tall slender individuals with poor posture hyperextend the lumbar regions of the spinal columns The long slender type of back is unable to withstand an increased strain for any length of time (Fig 6 1)

Alteration in Plane of Facets The frequency with which alteration of the plane of articular facets of the lumbar portion of the spinal column occurs is not known When in the three quarter roentgenographic view of the spinal column the facet or facets are found to be placed in a plane that is different from that of the other lumbar facets the joint cannot move in unison with the remaining joints of the lumbar part of the spinal column and thus function is impaired and irritative phenomena at the site of the anomalous articulation may produce pain

Infections of the Vertebrae The blood borne organisms find vertebral bodies and plates a suitable place to live The intervertebral disks are relatively avascular and are usually involved secondarily

Symptomatically the patient has a backache which is usually sudden in onset severe and disabling along with fever and leukocytosis Objectively the patient

the shaft of the femur (5) carcinoma of the rectum or of the prostate (6) malignant disease of the female genitalia (7) the pains of tabes or of dementia paralytica (8) ruptured intervertebral disk (9) intermittent claudication (10) neuritis of diabetic or other metabolic or nutritional causes and (11) metallic poisonings However backache with sciatic extension is most frequently associated with arthritic changes of the lumbosacral joints

Sciatica (*Sciatic Neuralgia*) Sciatica or neuralgia of the sciatic nerve is characterized by pain in the distribution of the sciatic nerve The causes enumerated in the list of diseases and conditions which may cause backache with sciatic pain are many (*vide supra*) Many of these diseases may be difficult to diagnose It is for these reasons that the diagnosis of sciatica without a cause is not safe to make However in practice there are a number of patients who will have sciatica from unknown causes the idiopathic sciaticas

These instances of the so called sciatic neuritis or ischemic sciatica (idiopathic sciatica) occur more frequently in men between the ages of 30 and 50 years of age than in women or than in men of other ages The symptoms commence with pain situated mainly in the back of the thigh The pain is dull and almost continuous with paroxysms of sharp pain and burning The nerve is tender on palpation

There are other neuralgias of the lower extremity for instance those of the femoral nerve (anterior crural neuralgia) the external cutaneous femoral nerve (meralgia paraesthetica) or the obturator (obturator neuralgia)

Meralgia Paraesthetica This is a variety of pain localized in the area of the distribution of the external cutaneous nerve on the lateral side of the thigh The pain is often induced by standing or walking Obese middle aged men are more commonly affected than are women The presence of an area of impaired sensation indicates a neuritis rather than a neuralgia Bilateral pain has been observed

Sciatic Scoliosis Ellis employs the term sciatic scoliosis to designate lateral deformity of the trunk secondary to a painful minor lesion in the lower part of the back the buttocks or the sciatic nerves The characteristic lateral tilt of the trunk represents a posture which will relieve or mitigate the pain producing the syndrome

The etiology of sciatic scoliosis is variable and may be difficult to elucidate When the cause is a lumbosacral pathologic change there is a rather sharp lateral deviation of the lumbar portion of the spinal column at the lumbosacral junction without any pelvic rotation

In spinal arthritis and sacro iliac disease there may be a generalized bending of the lumbar portion of the spinal column without rotation and deviation If bending of the lumbar vertebrae is great enough the scoliosis may be compensated for by a slight tip of the pelvis upward on the side toward the convexity and the leg on the affected side may be flexed at the hip and knee

A myelogram may be required to eliminate the possible presence of a protruded intervertebral disk

Traumatic Sciatica Woltman recorded 77 cases of neuritis of the lumbosacral plexus in each of which there was sciatic pain In 11 of these cases the neuritis was due to trauma such as excessive flexion of the thigh hyperextension of the thigh during a dive fracture of the sacrum football injury or a fall from a horse

During sleep a postural injury of the nerve may occur When the lower extremities are compressed in some unusual position as over the edge of a bed rail whether the pressure injures the nerve fibers or the nutrient vessels of the nerve trunk is conjectural

Other Forms of Backache Other less severe and some less common types of pain in the back that occur often enough and present enough similarities to justify mentioning are nocturnal backache the backache that accompanies menstruation and postmenopausal osteoporosis and the so called interspinous ligament syndrome

taken place A period of more than 7 weeks should arouse suspicion of superimposed disease such as arthritis or ruptured intervertebral disk, or of malingering

Results of the roentgenologic examination are negative

Muscle Rupture When a muscle ruptures a distinct gap can be felt between the retracted ends of the torn muscle and there is immediate disability consisting of local pain loss of power and swelling Such a condition does not often occur in the spinal muscles

Shoveler s Disease Wetzel directs attention to the occurrence of avulsions of the spinal processes in the course of exertions such as pitching hay in farm work, by persons who are unaccustomed to hard physical labor The complaint is pain between the shoulder blades There is sensitivity to pressure in this region and forward and sideward lifting of the arms causes pains The diagnosis is established by roentgenologic examination Such patients should refrain for about 4 weeks from the exertion that caused the avulsion

Degenerative Changes of the Vertebrae The lumbosacral region is affected by degenerative changes to a greater extent and at an earlier age than the other regions of the spinal column the cervical region being the next most susceptible

Degenerative changes cause most of the backache in the lower portion of the lumbar segment where stress is greatest Degenerative changes are designated as wear and tear changes osteoarthropathy bony spurring and narrowing of the intervertebral disk

Degenerative changes in the spinal column are not limited solely to the bones Degenerative changes occur in the skin the subcutaneous fat fascia muscles ligaments tendons bone and cartilage The muscles soon lose their elasticity and ability for rapid co ordination The ligaments surrounding and supporting the spinal column degenerate more rapidly than the muscles Changes in the vertebrae follow changes in the soft tissue

LUMBAR OSTEOARTHRITIS In arthritis of the joints of the lumbar portion of the spinal column pain may appear suddenly as in lumbago while the patient is stooping or in some disadvantageous position Certain positions in sitting are avoided unless the lumbar portion is supported The pain is intermittent and is characterized by acute exacerbations

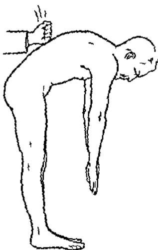


Fig 6 6 Percussion of the back

Examination usually reveals that the patient has evidence of lumbar joint disease and a superimposed myositis The myositis accounts for the acute exacerbation Local tenderness may exist on either side close to the spinous processes or in the interspinous ligaments Percussion may localize the soreness definitely (Fig 6 6) The roentgenogram may not corroborate the clinical findings since arthritis can seldom be visualized by the time the first symptoms appear

Backache and Pain in the Back With Sciatica Backaches may or may not be accompanied with extension of pain to the sciatic nerve and its distribution

The term sciatica describes a chronic and often remittent pain situated low in the back which extends down the course of the sciatic trunk in the back of the thigh The pain may extend to the distribution of the external peroneal nerve to the lateral surface of the calf and finally to the foot

Some of the causes of the symptom backache with sciatica can be enumerated thus (1) trauma (2) rheumatoid arthritis and osteoarthritis (3) spondylitis of the cervical or of the lumbar vertebrae and disease of the sacro iliac articulation (4) malignant disease of the pelvic bones or vertebrae or chronic osteomyelitis of

Nevertheless the findings are constant and characteristic enough so that a presump tive diagnosis can often be made before intraspinal roentgenographic studies are made

The diagnosis is made from properly interpreted spinograms Love and Walsh warn of the danger of missing a protrusion of the disk on roentgenologic examination after injection of radiopaque oil unless the lumbar portion of the spinal column is kept in extension

Malignant Metastasis to the Spinal Column Pain is the first symptom It is almost always limited to definite segments supplied by the nerve roots passing out ward through the foramina formed by the involved vertebrae The pain is aggra vated by coughing sneezing yawning straining at stool bending movements of the spinal column or sudden jarring of the body

The sufferer with metastatic lesions is reluctant to move or turn in bed because motion aggravates the pain Patients who have metastatic lesions of the spinal col umn are likely to walk the floor to avoid lying down Lying down often aggravates the pain

A common manifestation of metastasis of malignant disease to the spinal column is sciatica Any sciatic pain occurring in a middle aged patient either unilaterally or bilaterally which is not relieved by aspirin arouses the suspicion of spinal metastasis

A significant finding in those who have metastasis to bones is an elevated alka line phosphatase content of the blood serum In metastatic lesions of bone which result in increased localized calcification (increased osteoblastic activity) the serum phosphatase levels are almost always elevated With destructive (osteoclastic) lesions while the serum phosphatase values may be normal they are frequently elevated

The diagnosis is suspected from the history and the examination Roentgeno logic studies are of diagnostic value when two types of lesions are encountered (1) evidence of increased osteoblastic activity as indicated by localized areas of increased density due to increased calcification (2) localized rarefaction indicating increased osteolytic and osteoclastic activity with decalcification Increased concen trations of serum phosphatase are of diagnostic value

History of Backache It is essential to know (1) whether the pain in the back followed injury or unusual physical exertion (2) the exact location of the pain (3) whether the pain is continuous or recurrent (4) what makes the pain worse (5) what relieves the pain (6) whether the pain is getting worse and (7) whether a lawsuit is pending or anticipated It is unfortunate but nonetheless true that this last question has to be included If there is litigation pending (and frequently the patient is reluctant to admit it) it is essential that full details to include names places and time be obtained and be made a part of the written record

Patients who complain of disabilities associated with the back have pain of one of two types either static pain in the back or pain which recurs and lasts for periods of days or weeks with or without sciatic extension

Static pain the commonest of backaches is brought on by use or physical activity and it is relieved by rest Its presence denotes some fault in the moving parts of the back either posttraumatic or due to degenerative disease of the joints There may be a congenital anomaly or the back pain occurs in the absence of any demonstrable pathologic condition In these latter cases the pain is commonly due to poor postural attitudes which mechanically put undue stress on the ligaments muscles and small joints of the back Static type of pain is usually present daily and does not vary in intensity except as the intensity of the physical activity varies As a rule there are no severe acute episodes and there are no complete remissions Static pain in the back may at times be accompanied by some degree of fibrositis and the patient describes stiffness on arising jelling with inactivity or exacerbation

Of these backaches it is well to know that the backache which accompanies menstruation is rarely due to disease If it is due to disease at all the disease is intrinsic to the structures of the back or pelvis and proper examinations may detect the situation of the defective structures responsible for the pain

Injury Involving the Spinal Column When an injury of the *spinal column* has occurred it is important first of all to determine whether or not the spinal cord or the spinal nerve roots have been injured This necessitates a study of the motility the sensibility and the reflexes (deep superficial pupillary and sphincteral) Should a neural disturbance be found its exact site must be determined The site of the vertebral injury causing the symptoms can then be deduced from the interpretations by a roentgenologist

Protruded Intervertebral Disks In recent years it has been demonstrated that *protruded intervertebral disks* are common causes of intermittent backache The disks are ruptured by a sudden twist of the spinal column from a fall or by other injury of the spinal column Rupture of a disk may occur also as a part of degenerative disease of the vertebrae A disk may be ruptured or protrude in any region of the spinal column from the first cervical vertebra to the last lumbar vertebra However rupture most commonly occurs in the lumbar region

The symptoms are characterized by episodic backache which usually comes on during back motion The pain is often incapacitating for a week or 10 days and then subsides A recurrence of the pain is to be expected at some future date

During the attack coughing sneezing and blowing the nose or straining accentuates the pain in all instances During the interval between the attacks there is no distress During an acute episode the patient cannot stand erect or sit comfortably and the pain may be worse at night than during the day These patients may or may not have pain in the leg However pain is usually present in the leg and it may vary from a simple overflow of localized low back pain into the buttock to severe root pain that extends all the way down the leg into the toes There may be accompanying alterations in sensation and paresthesias at times Rarely there is some weakness or paralysis of peripheral muscles and more rarely some interference with function of intestines or bladder

In the standing position there is often a true sciatic and disk scoliosis which presents as a bend principally affecting the lumbar vertebrae and the shoulders are maintained at the same level by means of elevation of the pelvis on the affected side the body being thrown out of alinement toward the opposite side so as to maintain the correct shoulder level A fixed reversal of the lumbar curve with the most prominent part of this kyphosis at the third fourth and fifth vertebrae is present during an exacerbation of symptoms

Manipulation of the back in the sitting position may elicit little or no pain on rotation or lateral motion while the spinal column is flexed but when it is hyperextended and these motions are repeated all are painful Love and Walsh explain this symptom by their observation that the herniated disk is likely to be reduced by flexion of the lumbar portion of the spinal column whereas hyperextension on the *contrary* causes the disk to protrude more markedly

The Achilles reflex which subserves the fourth and fifth lumbar and first and second sacral segments of the spinal cord is lost or diminished in more than one half of cases of disks at the lumbosacral junction Protruded disks at the fourth interspace are found to reduce the Achilles reflex in some cases but protrusion at the third lumbar interspace decreases the ankle jerk in one half of these cases The patellar reflex which subserves the second third and fourth lumbar segments of the spinal cord is reduced or absent in one half of the cases in which a protruded disk is present at the third lumbar interspace

There are no physical neurologic or orthopedic signs which are found in all cases of protrusion of intervertebral disks that are characteristic of this injury only

MENSURATION Measurement of chest expansion is indicated when ankylosing spondylitis is suspected Measurement of the length of the legs may reveal a shortening which is influencing the spinal mechanics The calves should be measured at their greatest circumference and the thighs at comparable measured levels below the anterior superior iliac spines

The Sitting Position Have the patient sit on a stool without a back so that the feet rest comfortably and squarely on the floor Observe the lumbar region for evidence of whether or not the weight is borne equally on the two ischial tuberosities A manifest desire to sit on one ischial tuberosity and complaint of pain when asked to sit squarely on the stool arouse the suspicion that there is lumbosacral sacro iliac or coccygeal soreness

If the normal lordosis of standing persists while sitting the curvature may be at a higher level than usual because mobility of the lumbosacral joints is deficient or it may be at the lumbosacral junction as in spondylolisthesis

The Supine Position An examining table long enough for the patient to lie at full length or one adjustable to full length is better than a shorter higher table If a stool is used to climb to the table a patient with a unilateral back lesion will step up on the good leg

Ellis makes the following observations on patients as they get on the examining table A diffuse tenderness in the lumbar region *will* cause the patient to crawl onto the table in the prone position and then roll over with some difficulty A patient who has coccygodynia objects to lying flat on the table A patient who first sits on the examining table and then turns around so that a leg may be supported with his hands in bringing it up on the table is protecting a unilateral lesion on the side of the supported leg A lumbosacral or sacro iliac soreness may prevent lying flat on the back on the table Lying is more comfortab'e if the leg and back are rotated slightly toward the painless side The insistence on slight flexion of the hip while lying is more suggestive of sacro iliac than of lumbo sacral soreness Observe whether or not lordosis is changed by the patient flexing the hips and knees In lumbosacral soreness the lumbar lordosis does not change in sacro iliac soreness the lordosis is effaced

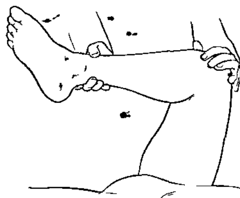


Fig 6 7 Internal rotation of hip joint is free of pain in the absence of disease of the hip

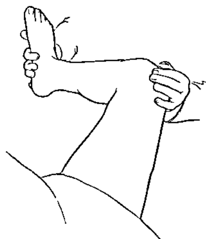


Fig 6 Lateral rotation of hip is f ee of pain in the absence of di ease of the hip

Application of pressure on the knee during this manipulation with production of pain is a positive Laguere s sign (see text)

during cold damp weather The fibrositis in these instances is secondary to the underlying condition responsible for the pain and is not the syndrome of primary fibrositis

The recurring or intermittent type of pain in the back comes on in acute attacks The attacks occur suddenly while the back is in motion The pain may be incapacitating may last from 5 to 10 days or more and may recur 2 or 3 times annually Such symptoms are commonly produced by protrusions of intervertebral disks tumors of the spinal cord spondylitis due to either tuberculosis or brucellosis benign lesions or metastatic neoplastic conditions of the vertebrae

Examination of the Back The patient is examined in the standing the sitting the supine and prone positions Inspection of the normal back has been described (p 186)

The Standing Position The examination of the back begins by desiring the patient to walk if walking is possible without too much pain The examiner observes the manner of station and gait The patient is then asked to sit down take off the shoes and stockings performance of these tasks also is observed The patient who has a painful back moves carefully and with evidence of stiffness

Usually the tilt of the trunk is away from the painful side in sciatic scoliosis but occasionally it may be toward that side Observe whether or not there are flattening or the lack of lumbar lordosis ankylosing spondylitis prominent spinous process of the fifth lumbar vertebra in spondylolisthesis angular kyphosis of Pott s disease rounded dorsal kyphosis of Scheuermann s disease and atrophy of the calf

MOTIONS OF THE SPINAL COLUMN The motions of the spinal column are extension flexion rotation and lateral bending The ranges of these motions are determined while the knees of the patient are held straight

A limitation in flexion of the spinal column in the standing position should be compared with the range of motion at which the pain appears when these same motions are performed when the patient is sitting normally upright

Lateral motion in the lumbar region is confined to an arc of not more than 40 degrees The extent of active and passive lateral motions is compared with this normal range The lateral motions of the lumbar vertebrae are better observed with the patient in the sitting position than in the standing position In the standing position spasm of the tendons bounding the popliteal space may cause a limitation of motion

Flexion and extension are free movements of the neck and lumbar regions Rotation is free at the articulation of the axis with the atlas It is limited in the mid cervical region freer in the upper portion of the thoracic region gradually diminishes downward and is absent in the lumbar region

The degree of referred pain if any on each motion is significant as an aid to the localization of disease On flexion the patient will stoop toward the side of the spastic erector spinae group of muscles which is ordinarily toward the side of the pain

Record should be made as to which of these motions if any reproduces the pain of which the patient complains In the matter of forward flexion one should distinguish between movement in the spinal column itself and movement accomplished by flexion at the hip joints

PALPATION Palpation and percussion over the spinous processes lumbosacral and sacro iliac joints should be carried out The amount of spasm of the paravertebral muscles is estimated One should ascertain whether or not there is tenderness on pressure over the sciatic notches or over the sciatic nerve itself in the thigh In fibrositis the demonstration of palpable tender nodules is diagnostic

PERCUSSION Percussion of the back is usually performed with the closed fist (Fig 6 6)

LASEGUE'S SIGN The (modified) straight leg raising sign of Lasègue is elicited by elevating the limb with the knee extended Approximately the first 20 degrees of this motion produces flexion of the hip and may produce pain in the hip joint As the motion continues and a pull is exerted on the ischial tuberosity by the hamstring muscles rotation is exerted on the sacro-iliac joint When the leg approaches a vertical position or points straight upward torsion increases rapidly and begins to be strongly transmitted through the lumbosacral joint of the same side At right angle flexion even soreness in the opposite lumbosacral joint will begin to be manifest

In sciatica if the straight leg test is performed to the point where pain begins sudden dorsal flexion of the ankle will greatly aggravate the symptoms (Fig 6 12)

LAGUERRE'S SIGN This sign is elicited by flexion abduction and outward rotation of the hip These movements force the femoral head against the anterior portion of the capsule and pain results if there is hip joint disease This test is used to distinguish psoas spasm hip joint disease and sacro iliac disease from lumbosacral disease (Fig 6-8)

GAENSLEN'S SIGN Gaenslen's test is used to elicit soreness in the sacro-iliac joint The opposite hip and knee from the side to be tested are fully flexed and the patient locks the lumbosacral and sacro-iliac joints on the opposite side by firmly encircling the knee with his arms The hip on the side to be tested is then hyperextended over the edge of the table with the knee flexed Pain is produced by rotation of the sacro-iliac joint of the hyperextended leg

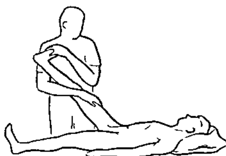


Fig 6 12 Lasegue's test Straight leg raising test with knee kept extended while pressure is applied to the foot



Fig 6 13 Sacro iliac tenderness demonstrated by downward pressure on the iliac crests

The legs are now placed close together with the knees extended and are both raised with one hand of the examiner It is the object of this maneuver to test the lower lumbar portion of the spinal column rapidly It has been observed that tension on the sacro iliac joints and the sciatic nerves is not produced until the hips are flexed through a considerable range so that when pain begins in the lumbosacral region as soon as flexion begins sooner than when either leg is raised separately the suspicion of disease rests on the lower lumbar portion of the spinal column and lumbosacral joints rather than on the sacro iliac joint and the sciatic nerve

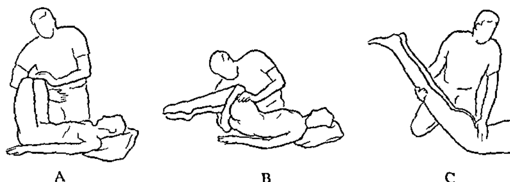
PATRICK'S SIGN Evidence of hip joint disease in which the pain is more likely to be referred along the anterior surface of the thigh than elsewhere is elicited by placing the ankle of the affected side above the patella of the opposite outstretched leg then pressing the flexed knee down

BAER'S SACRO-ILIAC POINT OF TENDERNESS Before concluding the examination in the position for eliciting Patrick's sign palpation should be made deeply in each lower quadrant of the abdomen to elicit Baer's sacro iliac point of tenderness which is described as situated 2 inches from the umbilicus on a line between the umbilicus and the anterior superior iliac spine Sacro iliac tenderness may also be elicited by downward pressure on sacro iliac spines (Fig 6 13)

The Prone Position When it is possible for the patient to lie in this position the examination should be continued as an inspection to estimate (1) the amount of lateral deviation as compared with that present in the standing and sitting positions if any exists and (2) the degree to which such distortion of the spinal column

Hip Motion When the knee is flexed motions of the hip joint can be studied without the transmission of tension to the sacro iliac or the lumbosacral joints Any complaint on internal or external rotation at the hip with the hip and knee flexed directs suspicion to the hip joint itself The motions of the hip joint may be tested as shown in Figures 6 7 and 6 8

Lateral and rotary motions of the lumbar portion of the spinal column while the hips and the knees are flexed are performed by the examiner pressing the knee forward laterally and downward as desired (Fig 6 9) Passive flexion of the lumbar



Modified from F B J D *The Injured Back and Its Treatment* Charles C Thomas 1940

Fig 6 9 A flexion and rotation of the hips may identify lumbosacral soreness as well as the extent of spasm or contraction in sciatic scoliosis In acute injury of a sacro iliac joint com pressive force (forceful flexion of both hips) is not so painful as axis traction or separation

B in lumbosacral joint disease strain on the sacro iliac joints exerted as shown elicits *tenderness in the lumbosacral joint*

C fixing the spinal column and hyperextending the legs permit observation of the position at which pain begins The force acts successively on the lumbosacral junction the sacro iliac joints and up the spinal column as the point of fixation is moved upward

joints can be performed through a larger arc with less pain in the supine position than in the standing or the sitting position Compression of the anterior superior spines of the ilia toward each other and separation from each other as *tests for* sacro iliac disease are performed in this position In sacro iliac disease these two maneuvers cause a complaint of pain

The straight leg raising tests are most valuable in detecting disease of the sacro iliac joint and disease of the lower lumbar region of the spinal column (Figs 6 10 and 6 11)

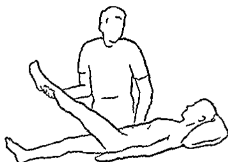


Fig 6 10 Straight leg raising test Hip free This much flexion at the hip is usually without pain when the hip is not diseased



Fig 6 11 Straight leg raising test with hip held down Less flexion at the hip is per missible without the production of pain when the hip is held down than when it is free

nates in the subclavian vein which lies to the inner (anterior) side of the artery

Immediately below the clavicle is a depression the infraclavicular fossa On the sternal side of this depression can be felt the first rib Since ribs are not always easily identified it is best in counting them to locate the angle of the sternum (angle of Ludwig) The second rib is immediately opposite this angle The angle of the sternum is about 2 inches (5 cm) below its upper edge Attached to the lower edge of the inner half of the clavicle is the pectoralis major muscle and to the outer third the deltoid muscle

Only a small part of the lower edge of the clavicle is free from muscular attachments This muscle free edge forms the base of the *subclavicular triangle* and its two sides are formed by the adjacent edges of the pectoralis major and deltoid muscles This triangle marks the course of the underlying first portion of the axillary artery with the vein and the cords of the brachial plexus of nerves Deep pressure appropriately applied here can compress the axillary artery against the second rib but not so effectively as above the middle third of the clavicle The pressure on the artery is immediately manifested by tingling along the inner side of the arm

Just to the outer side of the junction of the middle and outer thirds of the clavicle in front of the deepest part of the concavity is the coracoid process of the scapula It is best felt by pressing the fingers flat on the surface Running from the coracoid to the acromion process is the sharp edge of the coraco acromial ligament

Beneath the acromion process but covered by the deltoid is felt the greater tuberosity of the humerus If the arm is placed alongside the body with the palm facing forward a distinct groove can be felt to the inner side of the acromion process It is the bicipital groove of the humerus for the long tendon of the biceps muscle The greater tuberosity of the humerus projects beyond the acromion process and forms the prominence of the shoulder On rotating the arm the greater and lesser tuberosities of the humerus can be distinctly felt moving under the deltoid muscle

The acromion process of the scapula posteriorly forms a distinct angular prominence where it joins the spine of the scapula This prominent angle can be seen and felt and is used as a landmark for measuring the length of the humerus The spine of the scapula ends at the posterior border of the scapula This border is opposite the fourth rib and the third thoracic vertebra With the arm to the side the lower angle of the scapula lies over the seventh interspace

THE SHOULDER

Muscles The shoulder embraces two sets of muscles one set connects the shoulder girdle with the trunk and the other the humerus with the shoulder girdle

Movements The main movements of the shoulder girdle are anteroposterior as in swinging the arm those of abduction and adduction as in raising and lowering it sidewise and rotary

Functional Testing of the Muscles of the Shoulder *Deltoid* The examiner standing in front of the patient requests the patient to raise the arms laterally to a horizontal position Inability to do so indicates deltoid paralysis Also the deltoid muscle may be conveniently tested as shown in Figure 6 14 (Circumflex nerve supply C_5 C_6)

Pectoral The examiner stands in front while the patient stretches out the arms straight in front the examiner then approximates the patient s hands against resistance of the pectoral muscle Paralysis is indicated by failure of contraction

The two parts of the muscle are tested the sternocostal part as depicted in Figure 6 15 and the clavicular part as in Figure 6 16 (Lateral and medial pectoral nerve supply C_5 C_7)

Latissimus Dorsi The patient raises the arms laterally to a level then while keeping them fully extended brings them downward and backward as if to make

can be corrected after attempting to pull down the leg on the concave side of the lumbar curve

Roentgenograms No examination of the back is complete without antero posterior and lateral roentgenograms of good quality A localized lateral roentgenogram of the lumbosacral joint is necessary when a lesion at this site is suspected In fact when the nature of a lesion is doubtful localized anteroposterior and lateral roentgenograms of any part of the spinal column are indicated Oblique views are useful when one suspects a lesion of the articular facets or open neural arches in cases of spondylolysis and spondylolisthesis When a lesion of the sacro iliac joint is suspected stereoscopic anteroposterior roentgenograms of the pelvis will give the most information

Patients with ruptured intervertebral disks may require myelographic examina tion

SUMMARY OF BACKACHE

Many backaches are left unexplained after all examinations have failed to reveal disease Most persons who have these unexplained backaches continue to live normal active lives despite the back pain

Perhaps the two commonest ailments to cause soreness and pain in the back are *fibrositis and psychoneurosis In women backache is rarely due to disease of the* pelvic organs

In a patient known to have malignant disease an unexplained backache espe *cially if there is sciatic distribution of the pain or extension of pain to the leg* excites the suspicion that the cause of the pain is metastasis to the spinal column

In a middle aged person the onset of a backache with sciatic distribution which is not relieved by aspirin excites the suspicion that there is metastasis to the spinal column from an unknown malignant source The suspicion is strengthened if there is bilateral distribution of the pain

THE APPENDICULAR SKELETON AND LOCOMOTION

THE SHOULDER GIRDLE (CINGULUM EXTREMITATIS SUPERIORIS) AND THE UPPER EXTREMITY

Topography and Bones The shoulder girdle projects the arm well out from the trunk by means of a framework which consists of three bones the clavicle and scapula superiorly and the humerus inferiorly These bones form the basis of the shape and topography of the shoulder The shape may be modified by the muscles and fat

The clavicle is subcutaneous and can be felt throughout its length The medial end normally projects above the upper edge of the sternum which can be felt at *the suprasternal notch A prominent ridge marks the outer extremity* of the clavicle if this ridge is difficult to recognize as is often the case the examiner continues directly laterally to the point of the shoulder which is formed by the tip of the *acromion process This point having been recognized the end of the clavicle* is found directly mediad from it

In the median line above the sternum is the suprasternal notch Just to each lateral edge of the notch is the *prominent sternoclavicular joint* On the right side this joint marks the ending of the innominate artery and the commencement of the right common carotid and subclavian arteries That on the left marks the left carotid with the subclavian *directly to its outer side and a little posteriorly*

The medial two thirds of the clavicle is convex anteriorly The lateral third of the clavicle is convex posteriorly and from its superior surface the trapezius muscle can be felt *Just above the clavicle in the neck and behind the clavicular* origin of the sternocleidomastoid muscle is seen the external jugular vein It termi

scapulae as close together as possible (Fig 6 21) (Accessory nerve supply C_3 C_4)

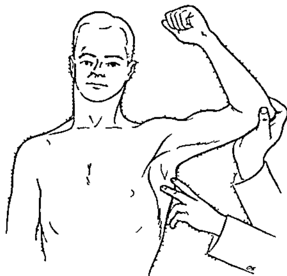


Fig 6 17 Latissimus dorsi muscle (insertion) Standing or sitting with the arm horizontally and laterally extended Attempts are made to extend the arm against resistance The muscle can be seen and palpated with the middle finger Nerve supply C_6 C C_8

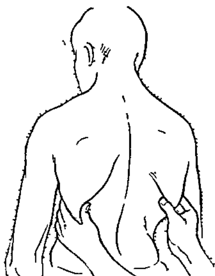


Fig 6 18 Latissimus dorsi muscle (posterior portion) Standing or sitting On coughing the muscle can be felt except in extremely obese persons Nerve supply C_6 C_7 C_8

Rhomboideus Muscles The patient moves the shoulder backward against resistance (Fig 6 22) (Nerve supply C_4 C_5)

Supraspinatus Muscle Standing with the arm by the side attempts are made to abduct the arm against resistance The muscle can be felt on top of the shoulder (Fig 6 23A) (Suprascapular nerve supply C_5)

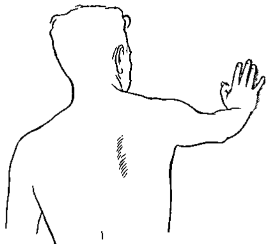


Fig 6 19 Serratus anterior muscle While standing with hand against wall patient pushes forward The medial border of the scapula remains close to the thoracic wall which can be seen or felt Nerve supply C_5 C_6 C_7

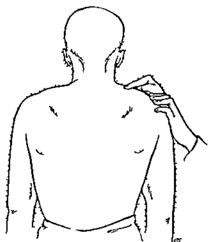


Fig 6 20 Trapezius muscle (upper part) Standing or sitting Attempts are made to elevate the shoulder against resistance The midportion of the muscle can be seen and felt Nerve supply C_3 C_4 (accessory nerve eleventh cranial)

the hands meet behind the sacrum The examiner stands behind the patient to resist the movement The muscles are also conveniently tested as the patient tries to adduct the horizontally and laterally extended arms against resistance (Fig 6 17) If the

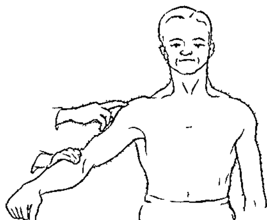


Fig 6 14 Deltoid muscle Standing or sitting Attempts are made to maintain an angle of abduction of the arm of more than 15 degrees against resistance The muscle can be seen and felt Nerve supply C C_6 (circumflex nerve)

patient coughs the muscle bellies can sometimes be seen and palpated (Fig 6 18) (Nerve supply C_6 C C_8)

Serratus Anterior Request the patient to push with his hands against the wall If the serratus has lost its power the scapula will project and the digitations of the muscle which should be visible will not be seen (Fig 6 19) (Nerve supply C C_6 C)

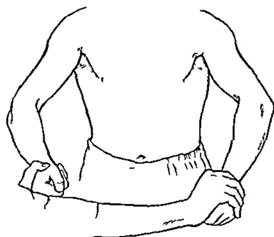


Fig 6 15 Pectoral muscles (sternocostal portion) The patient standing or sitting The arms are raised below the horizontal Attempts are made to abduct the arms against resistance The sternocostal position can be seen and felt Nerve supply C_6 C_7 C_8 T_1 (lateral and medial pectoral nerves)



Fig 6 16 Pectoral muscles (clavicular portion) Standing or sitting The arm is raised above the horizontal and forward flexed at the elbow Attempts are made against resistance to abduct the arm The clavicular and usually the costal parts of the muscle are evident Nerve supply C C_6 C_7 C_8 (lateral and medial pectoral nerves)

Trapezius The examiner asks the patient to raise the shoulder upward against the pressure of the examiner s hand This will demonstrate the strength of the upper part of the trapezius (Fig 6 20) The examiner asks the patient to bring the

other causes for shoulder pain Some of these causes are intrinsic while others are extrinsic and these may be enumerated as (1) arthritis (rheumatoid and osteoarthritic tuberculous and due to pyogenic infections) (2) tumors and infections of the scapula or head of the humerus (3) lesions of the clavicle (4) lesions of the acromioclavicular joint (5) tumors of the soft tissues about the shoulder (6) fractures or fracture dislocations of the shoulder and (7) diseases of the diaphragm which often cause pain in the shoulder

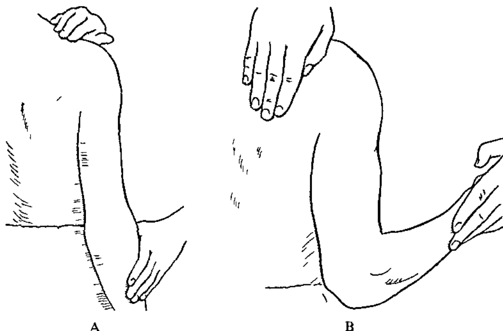


Fig 6 23 A supraspinatus muscle Standing with arm by the side attempts are made to abduct the arm against resistance The muscle can be felt on top of the shoulder Nerve supply C (suprascapular nerve)

B infraspinatus muscle Standing or sitting with the elbow flexed to the side resistance is made against attempts to rotate the arm externally at the shoulder joint The muscle can be felt Nerve supply C C_6 (suprascapular nerve)

The differential diagnosis of lesions of the shoulder from lesions of the cervical portion of the spinal column or brachial plexus often is difficult It is recalled that in cases of pain in the shoulder or arm lesions of the cervical part of the spinal column and shoulder joint may be present The most important point in differential diagnosis is the presence of local signs in the shoulder that is both active and passive limitation of motion either diffuse or localized tenderness atrophy of variable severity and swelling or local heat in an occasional case of acute bursitis In cases in which the shoulder joint is free of all of these signs the primary cause of the trouble usually is elsewhere The pain is referred pain

A number of lesions of the thorax and a few of the abdomen which irritate the diaphragm may cause shoulder and arm pain Of these cardiac pain may be referred to the arm first According to H L Smith the most important feature of such pain is that it must be produced by effort It occurs during effort and not afterward

Lesions of the superior pulmonary sulcus adjacent to the brachial plexus or sympathetic chain also may cause pain of the arm Pancoast described a syndrome including roentgenographic shadows of the tumor at the apex of the lung a neuritic type of pain of the arm atrophy of the muscles of the hand and arm and Horner s syndrome The *Pancoast syndrome* is not a distinct clinical entity since neuro-

Infraspinatus Muscle Standing or sitting with the elbow flexed to the side resistance is made against attempts to rotate the arm externally at the shoulder joint The muscle can be felt (Fig 6 23B) (Suprascapular nerve supply C C₆)

MONARTICULAR DISORDERS

These affect intra or extra articularly, a single shoulder, elbow hip or knee

The Shoulder and Monarticular Disorders The etiologies of the monarticular involvements the syndrome or syndromes of arm pain and their relation to lesions of the cervical part of the spinal column shoulder and brachial plexus are not always possible to delineate and to elucidate

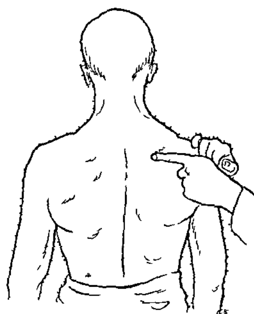


Fig 6 21 Trapezius muscle (adductor scapulae portion) Standing or sitting At tempts are made to carry the shoulder backward against resistance The lower part of the muscle can be felt and often seen Nerve supply C_3 C_4

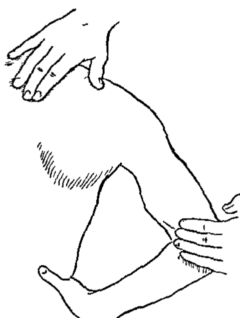


Fig 6 22 Rhomboideus muscles Patient standing elbow flexed and extending laterally with hand on hip tries against resistance to brace shoulder backward The muscle can be seen and felt Nerve supply C_4 C_5

The painful lesions of the shoulder due to a slowly developing internal derangement some of which are associated with trauma others not are (1) frozen shoulder capsulitis tendinitis peritendinitis or periarthritis (2) calcified subdeltoid bursitis or degeneration of the supraspinatus tendon and (3) tears of the supraspinatus tendon and of the musculotendinous cuff Regarding these painful shoulders certain facts as enumerated by Ghormley have come to be accepted 1 The supraspinatus tendon tends to degenerate and the frequency of this occurrence increases with age 2 There is general agreement that the so called periarthritis or frozen shoulder has its inception in inflammation of the tendon or tendon sheath 3 Calcification in bursae probably results from accumulation of degenerated tendinous material 4 Ruptures of the supraspinatus tendon are fairly common in older persons They may cause severe symptoms in some cases and require surgical treatment 5 Among younger persons subsequent to trauma tears of the musculotendinous cuff are much more severe and serious than among older persons

In addition to these facts enumerated by Ghormley Bickel has enumerated

with hypesthesia paresthesia or anesthesia In time atrophy of the muscles of the hands supervenes If the pressure and circulatory insufficiency are allowed to con tinue pressure on the subclavian artery decreases the volume of the radial pulse Peripheral thrombosis may occur and may be so extensive that gangrene ensues

EXAMINATION Palpation of the supraclavicular fossa is made in an attempt to detect a *cervical rib or any difference in the prominence of the structures in the* fossa and tenderness on pressure over the scalenus anticus muscle Pressure over this muscle causes extension of the pain down the ulnar side of the arm

The two radial pulses are examined for evidence of differences in the volumes While palpating the radial pulse request the patient to turn the head upward toward the affected side to inspire and to hold the breath If there is abnormal pressure by the scalenus anticus muscle on the artery the pulse volume will be decreased or the pulse may be obliterated and the symptoms will be reproduced

DIAGNOSIS The diagnosis of the scalenus anticus syndrome is based on the pres ence of pain atrophy circulatory abnormalities and disturbance of sensation in the *upper extremity The presence of cervical ribs is not sufficient evidence to warrant* a diagnosis of the scalenus anticus syndrome since as stated cervical ribs usually do not present symptoms

DIFFERENTIAL DIAGNOSIS The differential diagnosis of scalenus anticus syn drome requires a consideration of the following (1) brachial neuritis (2) Ray naud s disease (3) thromboangiitis obliterans (Buerger s disease) (4) subacromial bursitis (5) protrusion of a cervical intervertebral disk and neoplasms of the cervical portion of the spinal cord and (6) cervical spondylitis

THE AXILLA

Have the patient raise the arm directly out from the body The armpit and axillary folds are thus made visible

The axilla is a wedge shaped space with its apex upward formed between the arm and chest at their junction It serves as a passage for the arteries veins nerves and lymphatics between the trunk and the upper extremity It is frequently the site of growths and abscesses

The rounded edge of the anterior axillary fold is formed by the pectoralis major muscle It follows the fifth rib and its upper end merges with the lower edge of the deltoid muscle The posterior fold is formed by the latissimus dorsi and teres major muscles By deep upward pressure with the extended approximated fingers in the axilla the rounded head of the humerus can be felt

Lymphatic structures of the axilla especially the axillary lymph nodes combined with a few scattered nodes in the antebrachium and the brachial regions drain the entire upper extremity The axillary nodes receive the afferent vessels from the anterior and the lateral aspects of the thoracic wall from the breast and posteriorly from the scapular region These axillary nodes are anatomically situated in two groups the axillary nodes proper and the subclavian nodes

The lymphatics of the middle and lower portions of the back as low down as the umbilicus also drain into the axilla

These lymph nodes communicate with one another The part affected ordinarily drains to the nearest nodes Occasionally however the infection may pass by or through one set of nodes without causing enlargements and may involve a neighboring communi cating set *It happens sometimes in carcinoma of the* breast that the posterior or scapular set is involved and the anterior or thoracic set escapes

When the axillary lymph nodes are enlarged they may be seen and are always palpable When they are not enlarged they cannot all be found The size of the normal lymph nodes is usually inversely proportional to the number of nodes present In an axilla in which there are few nodes some of these may be palpable normally

The subclavian nodes lie in the infraclavicular triangle between the pectoralis major

fibromas or inflammatory lesions as well as malignant lesions may cause it Pain from lesions of the esophagus is rarely referred to the arm Diaphragmatic lesions however may cause pain in the arm This is particularly true of lesions of the central portion of the diaphragm such as esophageal hiatal hernias Diaphragmatic pleurisy or irritation also may cause pain in the shoulder and arm

Ankylosis of the Shoulder The relative loss of co ordination endurance and security in respect to the duties of the arm and hand will vary according to the extent the ankylosis approaches or departs from the more favorable position of about 50 to 60 degrees abduction (McBride)

Partial Ankylosis Partial limitation of motion in the shoulder is often due to a periarticular derangement of tissues resulting from trauma such as contusion and fractures of the head or surgical neck of the humerus A sprain or dislocation may leave permanent damage to the capsule and ligaments of the joint Habitual disloca tion is not commonly associated with a painful or stiffened joint The supraspinatus muscle or tendon may be ruptured and if so *the rupture is difficult to recognize*

Continuous immobilization with the arm to the side will result in permanent contraction of the adductors and atrophy of the deltoid so that there is no power to lift the arm outward Paralysis of the deltoid may follow a fall directly on the shoulder which injures the circumflex nerve Such a paralysis may be permanent

Suppuration and persistent sinuses may exist for many months after a compound fracture of the shoulder Movement of the joint will be greatly hampered Osteo *myelitis may follow a compound fracture and may cause permanent disability* Adhesions from this source do not respond well to manipulation

A rheumatic fibrositis may complicate injury to the shoulder When it does the pain is persistent and improvement is slow A deltoid bursitis may occur spon taneously or be induced by injury There may be a tenosynovitis of the long head of the biceps to add more to the pain and disability

When the scapulohumeral joint is ankylosed shoulder joint action is completely lost *if there is no movement of the scapula Ankylosis is incomplete* if the scapula is free With fixation of the arm at the side there is complete loss of motion The hand can perform its functions in the sagittal plane only by means of the forearm and the center of action is now situated at the waist instead of in the shoulder

When the scapula is free there is considerable freedom of motion rendered through the gliding of the scapula and through the sternoclavicular joint The amount of motion afforded through the scapula greatly lessens the severity of rigid ankylosis in the scapulohumeral joint

When fixation of the shoulder joint occurs with the arm more than one half abducted and slightly flexed with rotation at midpronation it is in a useful position This position permits more flexibility at the shoulder while lifting than other posi tions permit When the scapula is fixed the disability is much more severe than when *it is free*

The Scalenus Anticus Syndrome With and Without Cervical Rib The scalenus anticus syndrome may be associated with a cervical rib or it may occur spontaneously The symptoms are attributable to irritation and compression of the brachial plexus or the subclavian artery or to a combination of nervous and vascular symptoms However cervical ribs are of common occurrence and *usually do not* produce symptoms These ribs occur unilaterally or bilaterally and vary greatly in length

SYMPTOMS The commonest and often the first symptom is a sharp pain origi nating from a sudden rotation of the head or from a forceful downward *pull of the* shoulder In the intervals between these sharp pains there may be a dull aching burning and boring pain occurring during the latter part of the day or with fatigue The pain extends the length of the inner side of the arm and may be associated

Projecting on each side of the elbow are the two epicondyles of the humerus These bony projections do not belong to the forearm The two epicondyles are nearly on the same level The medial (internal) is the more prominent By deep pressure the lateral (external) supracondylar ridge can be felt running up the arm somewhat posteriorly from the lateral (external) epicondyle The medial (internal) supracondylar ridge is much less easily felt When the elbow is flexed at a right angle a plane parallel with the humerus passes through the two epicondyles and the tip of the olecranon If the forearm is extended the olecranon passes somewhat in front of this plane Hence in examining the elbow for injury it is desirable to determine the relation of these points when the elbow is bent at a right angle

The elbow is now examined while the arm is extended In this position the tip of the olecranon can be felt with the medial (inner) epicondyle to its inner side Between the medial and lateral epicondyles is a deep groove and in this groove can be felt the head of the radius

On the posterior aspect of the elbow joint is the ulnar nerve A bursa lies between the upper or posterior surface of the olecranon and the skin and also another on its inferior surface extending downward which from its exposed position is frequently injured and enlarged Such an enlargement occurs from chronic irritation in certain occupations (as mining) hence the name miner s elbow

In the middle of the flexor surface of the elbow below the crease is a depression called the antecubital fossa To its outer side is the muscular prominence of the extensors and supinator To its inner side is the muscular prominence of the flexors and pronator The inner muscular swell ends at the medial (internal) condyle but the external one passes well up on the arm

The *antecubital fossa* or space is occupied by a number of *veins* which are of importance because they are frequently used for purposes of parenteral medication sometimes for blood transfusions and not infrequently they are wounded and give rise to hemorrhage

The Elbow and Monarticular Disorders The diseases of the region of the elbow include inflammatory processes and neoplasms Inflammations of the elbow and its neighborhood may be acute or chronic In *acute arthritis* the whole circumference of the joint is tender on pressure the swelling being most marked where the capsule is most superficial

Chronic arthritis of the elbow joint may be due to rheumatoid arthritis tuberculosis or syphilis An arthritic involvement is not likely to be confused with simple chronic bursitis olecrani or with epicondylar neuralgia

Of the *tumors* of the region of the elbow those of intramuscular origin include angioma and sarcoma The simple muscle hernia due to the bulging of contracted muscle through a defect in the aponeurosis of a muscle following trauma or congenital defect is easily distinguished from neoplasm Similarly calcification within a *muscle following trauma the so-called traumatic osteoma in reality* a circumscribed ossifying myositis is differentiated from tumor by the history of trauma and by roentgenographic examination the appearance in roentgenograms is distinctly characteristic

Tumors originating in the nerves rather than in the muscles can be differentiated from muscle tumors by their position in the course of the nerves and by their spindle shape They consist usually of neurofibromas more rarely of sarcomas

Inflammations of the bones may be acute (osteomyelitis) or chronic (chronic staphylococcal osteomyelitis sporotrichosis tuberculosis or gumma) The neoplasms of the bones found in the region of the elbow are most often myeloid sarcomas occasionally other forms of sarcoma fibroma or enchondroma may be met with

Ankylosis of the Elbow The function of the elbow is to serve the hand and

and deltoid muscles and on the front of the subclavian vein above the pectoralis minor muscle They receive radicles from the mammary gland as well as from the axillary groups They receive the overflow either directly or indirectly from all the other lymphatics of the foregoing regions *They may empty through the subclavian trunk* into the subclavian vein near its junction with the external jugular vein or into the right jugular vein These nodes may be palpable in the presence of carcinoma of the breast

Axillary Tumors Tumors of the axilla are almost always due to involvement of the lymph nodes They may be inflammatory forming the ordinary axillary adenitis they may be tuberculous or they may be malignant Since the tumors of the axilla are often due to disease of the lymph nodes the parts which the nodes drain should be searched to determine the starting point of the disorder

Chronic infections of the glands of the skin cause tender axillary indurations Lipomas accessory breasts or breast tissues may be present in the axilla but more commonly these occur along its anterior margins

THE ARM

The arm has four sets of muscles to perform its four directional movements These sets of muscles are (1) the lateral group (abductors) (2) the internal group (adductors) (3) the anterior group (flexors) and (4) the posterior group (ex *tensors*)

The deltoid forms the large rounded prominence of the shoulder At its inser tion the humerus is nearest the surface and easily palpable The posterior edge of the deltoid can be plainly seen when contracted running upward and inward and crossing the posterior fold of the axilla at right angles Its anterior edge blends more or less completely with the pectoralis major

The biceps brachii and the brachialis form the muscular mass on the *anterior* surface of the arm The *posterior* or extensor set of muscles includes the triceps

On the medial aspect in the lower third of the arm the brachial artery is covered only by the skin and superficial and deep fascia and can be felt pulsating along the inner edge of the biceps muscle and tendon it can be compressed against the bone by pressure directed outward At this point tourniquets may be applied

THE ELBOW JOINT

The elbow joint is a hinge joint The articulation between the trochlea and the ulna is so shaped as to allow no lateral motion but only an anteroposterior one The forearm can be extended to an angle of 180 degrees or in a straight line with the arm It can be flexed to an angle of 30 to 40 degrees Sometimes it cannot be flexed so much so that if after an injury to the joint the patient can flex the elbow to half a right angle this may be regarded as the normal amount of motion

The axis of motion of the elbow joint (the radial aspect anteriorly) slopes slightly from the lateral side downward and inward The effect of this slope is to give a slight obliquity to the motions of flexion and extension This obliquity is not noticeable except in extreme extension and flexion When the forearm is completely extended (palm forward) it is seen not to lie in the axis of the arm but to bend outward from the elbow at an angle of 170 degrees This angle is called the *carrying angle* because by resting the elbow against the side any article which is carried in the hand as a bucket of water is kept away from the lateral aspect of the leg

In *examining the elbow* it first should be flexed at approximately a right angle The prominent olecranon process is the posterior projection of the elbow It is sub cutaneous and it can be felt and followed down the back of the forearm From the tip upward for a comparatively short distance can be felt the upper surface of the olecranon into which the triceps inserts To feel this upper surface distinctly the *forearm* should be somewhat extended to relax the triceps so that the outline of the upper portion of the olecranon is distinctly palpated

ample the screw driver) These instruments are therefore designed for right handed individuals A left handed individual s greatest handicap is that of having to use instruments which were not designed for him

THE WRIST

There is a bony prominence on each side of the wrist caused by the expanded lower end of the radius on the side of the thumb (medial) and head of the ulna on the side of the little finger (lateral) The medial (inner) prominence is exemplified by abducting the hand the lateral (outer) prominence of adducting it

Proximal to the wrist on the anterior and medial part can be felt the sharp and prominent distal 1 inch (about 2 5 cm) of the radius Following the bone distally on its medial side one feels the tip of the styloid process a most important land mark The styloid process is in the anatomist s snuffbox (tabatiere anatomique) a depression on the dorsal aspect of the wrist just below the radius

Between the tip of the styloid process of the radius and its sharp anterior border are the extensor ossis metacarpi pollicis and extensor brevis pollicis tendons The sheaths of these tendons frequently become inflamed from injuries If the examining hand is placed firmly on the lower portion of the radius of a patient so affected and the patient moves the thumb a characteristic grating irregularity of movement of the tendons can be palpated as they move in their inflamed sheaths

If the posterior surface of the ulna is followed downward the styloid process forming its extremity can be distinctly felt especially if the patient s hand is placed in the supine position and slightly flexed

The styloid process of the radius extends lower than that of the ulna Across the central (palmar) surface of the wrist there are two transverse lines The proximal or upper one corresponds to the wrist joint The distal or lower one corresponds to the joint between the two rows of carpal bones

On the dorsal surface of the wrist when in pronation one third of the width of the wrist across from the edge of the radius can be felt a bony prominence This prominence marks the middle of the posterior surface of the radius The radius occupies two thirds of the dorsal surface of the wrist and the ulna the other third by firm pressure the interval between the bones can be felt

The wrist joint has no movement of rotation Rotation of the hand is accomplished by pronation or supination of the forearm The hand can be flexed and extended through an arc of approximately 140 degrees and adducted and abducted about 70 degrees Adduction or bending toward the ulnar side is much greater than abduction or move ment toward the radial side

The extent of the movements of the wrist varies individually

Ankylosis of the Wrist The function of the wrist is to serve the fingers and hand in carrying out such duties as reaching pointing grasping pinching holding pushing pulling striking carrying swinging throwing turning and lifting The hand and fingers in execution and achievement of their appointed functions require the elbow and shoulder to serve their purposes

The causes of partial or complete ankylosis of the wrist are contusion or sprain tenosynovitis or ganglion fracture and dislocation suppuration continuous and pro longed immobilization scar contraction and shortening of muscles paralysis and rheu matic fibrositis

Partial Ankylosis Sprains are often accompanied by prolonged disability and may result in permanent loss of function Effusion into the wrist joint often hampers the joint motion for long periods of time if not permanently Sprains may be dis tinguished from tenosynovitis by the limitation of the swelling at the joint lines and do not extend up and down in the direction of the tendons In traumatic synovitis there is an aseptic effusion of blood and serum into the joint and in case of the wrist joint there are so many pouches and cartilaginous reflections about the carpal

arm in carrying out the motions entailed by reaching pointing holding pushing pulling striking carrying swinging throwing turning and lifting

Ankylosis of the elbow joint may result from dislocations contusions suppurations prolonged immobilization scar contracture myositis ossificans and rheumatic fibrosis

Partial Ankylosis When the range of elbow motion is less than a right angle the arm which is subservient to the hand does not provide flexibility Any motion at all is preferable to rigid fixation motion from 90 degrees to 35 degrees flexion is useful in the lighter activities but such motion will not permit carrying pulling or pushing heavy objects

When the range of motion of the elbow joint is from 180 to 80 degrees the greater portion of the requirements for accommodation of the hand and arm are fulfilled In the performance of the motions required for lifting pushing pulling or carrying the angle approaching 120 degrees is more desirable than a right angle (McBride)

When the elbow joint action is lost the arm is stiffened solidly The stiffened arm and the hand have their fulcrum at the shoulder and can perform only as far as the shoulder muscles are able to manipulate the entire arm The manual functions are greatly reduced because of inability to get the hand where it is needed

THE FOREARM

Anteriorly the muscles and tendons can be palpated The extent to which these can be outlined by the examiner depends on the absence of subcutaneous fat and the degree of development and contraction of the individual muscles The skin of the forearm is loose and thin The median vein is situated on the flat surface the radial vein winds around the back of the wrist and crosses the outer edge of the radius about its middle On the medial side near the elbow the anterior and posterior ulnar veins are visible as they pass posteriorly

On the posterior surface of the forearm the bones are more conspicuous than on the anterior surface because they are subcutaneous By palpation the ulna can be traced down the forearm running from the olecranon process down to its styloid process at the back of the wrist To the outer side of the olecranon can be felt the lateral (external) epicondyle and the capitulum humeri If the elbow is extended a dimple is seen just below the capitulum humeri it marks the position of the head of the radius

By placing the thumb of one hand in the dimple on the head of the radius and *rotating the hand of the patient with the other the examiner can feel rotation* of the bone and thus be assured that the radius is intact Whenever fracture of the radius is suspected this is the procedure resorted to in order to determine whether or not the radius is broken

The radius can be followed distally only for a distance of about 1 inch (about 2 5 cm) where it disappears beneath the muscles to become subcutaneous again on the lateral side of the middle of the forearm Thence it can be palpated to the styloid process on the lateral side of the wrist

Movements of the Forearm In the midposition with the radius above the ulna the space between the bones is at its maximum It is this space which if encroached on by poor apposition of fractured bones results in limitation of movements of pronation and supination The head of the radius rotates in the orbicular *ligament* the lower end of the radius revolves around the head of the ulna The range of movement is from 140 degrees to 160 degrees

The muscles of supination are stronger than those of pronation and it is for this reason that *instruments* intended to be used in a rotary manner are designed *to turn* from the inside toward the outside that is in the direction of supination (for ex

situated in the hand itself An especial accomplishment of the human hand is the ability to oppose the thumb to the other digits

Movements The motion between the individual carpal bones is limited to a slight gliding on one another The summation of these motions however permits considerable range of movements These movements are flexion and extension abduction and adduction and circumduction If motion is prevented in the bones of the forearm it is impossible to rotate the hand

The *radiocarpal joint* has a greater degree of flexion than of extension The movement permitted between the two rows of carpal bones is extensive

The movements of the four midcarpometacarpal joints are flexion (toward the palmar surface) and slight extension There is slight lateral flexion and extension which enables cupping of the hand for grasping round objects The articulation of the thumb allows flexion extension abduction adduction and circumduction but not rotation

The *metacarpophalangeal articulations* of the fingers are practically saddle shaped joints resembling somewhat the ball and socket joints with all their movements except that of rotation They can be flexed to an angle of 90 degrees The interphalangeal joints are hinge joints and capable only of flexion and extension The second joint can be flexed to an angle of 120 degrees and the end joint to about a right angle The prominence of the knuckle is formed by metacarpal bone

Functional Testing of Muscles of the Arm and Hand *Biceps* The patient extends the arm and flexes it at the elbow Further attempts of flexion are resisted by the examiner *The muscle can be seen and felt* (Fig 6 24) (Musculocutaneous nerve supply C C_6)

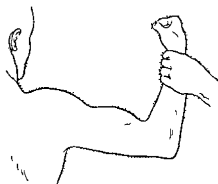


Fig 6 24 Biceps muscle Standing or sitting with the arm extended laterally *at the shoulder and supinated* and flexed at the elbow Attempts are made to flex the supinated arm at the elbow The muscle can be seen and felt Nerve supply C C_6 *(musculocutaneous nerve)*

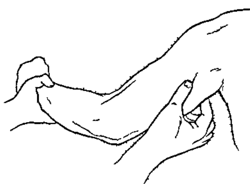


Fig 6 25 Triceps muscle Long heads Standing sitting or lying so that the force of gravity is inoperative *on the flexed arm at the* elbow Attempts are made to extend the fore arm against resistance The muscle can be seen and felt in either place shown Nerve supply C_7 C_8 *(radial nerve)*

Triceps The triceps may be tested as is the biceps except that the previously flexed arm is to be extended against resistance The muscle can be seen and felt The long heads of the triceps muscle can be palpated here (Fig 6 25) The whole or cross section of the triceps muscle can be palpated at the level of the arm indicated (Fig 6 26) (Radial nerve supply C C_8)

Brachioradialis With the forearm midway between pronation and supination the attempt at flexion renders the belly of the muscle evident (Fig 6 27) (Radial nerve supply C_5 C_6)

Extensor Carpi Radialis Longus With the fingers extended attempt is made to extend the wrist to the radial side The contracted muscle can be felt (Fig 6 28) (Radial nerve supply C C)

bones that adhesions from the coagula may become very extensive Rheumatic fibrositis is recognized by persistent and painful stiffness It is commoner in those past 40 years of age than in younger persons

As the angle of extension is lost in the wrist the flexion power of the fingers is diminished accordingly and the deformity becomes more disabling The prominence of the base of the hand over which the flexor tendons pass increases the flexor tension as the hand is dorsiflexed When the hand cannot be dorsiflexed to or past the 180 degree line therefore the long flexor tendons are at a disadvantage Rigid fixation in flexion eliminates the action of all muscles of the wrist The flexion power of the fingers is less when the fixed position is in abduction to the radial side than when in adduction to the ulnar side

When the wrist is fixed in *dorsiflexion* the palm of the hand is serviceable and the prominence of the base of the hand retains its function The flexor action of the fingers approaches normal The degree of adduction decreases the strength of fingers while abduction strengthens them The thumb flexors are improved in strength in the position of ulnar flexion or adduction In fixed ankylosis therefore midpronation is the position of choice

When the limitation of rotation originates at the elbow the biceps may be influenced through limitation of flexion

In the ankylosed position of full supination the hand is turned the wrong way to be useful As the range of motion approaches midway between pronation and supination the fingers and hand increase in their usefulness but the most useful position is that of about 35 to 45 degrees of pronation (McBride)

THE HAND

Man alone has a hand All other mammals have paws or front feet With respect to length the hand is classified is being long or midlong or short With respect to breadth the hand is spoken of as being broad midbroad or narrow Absolute breadth of the hand increases with age and is enhanced by physical labor in the early period of life so that among middle aged adults there are more than twice as many broad hands as all other widths combined

On the dorsum of the hand the extensor tendons can be seen Accessory slips usually connect the tendon of the ring finger with the tendons of the little finger and middle finger A slip also usually passes from the tendon of the middle finger to that of the index finger

On the dorsum of the hand the metacarpal bones are subcutaneous and can be felt in their entire length The muscular prominence on the dorsum of the hand seen when the thumb and the forefinger are approximated is due to the abductor indicis muscle At its upper extremity the radial artery passes between the two heads of the muscle to enter the palm When the thumb is extended the *anatomist s snuffbox* becomes evident and the extensor pollicis longus tendon is distinctly seen leading to the ulnar side of the posterior radial (thecal) tubercle on the middle of the dorsum of the radius The tendons on the radial side of the snuffbox are the extensor brevis and extensor ossis metacarpi pollicis

If the hand is turned with the palm up the thumb diverges from the median line at varying angles The palm is the hollow of the hand formed by a muscular mass on each side On the thumb side is the *thenar eminence* on the ulnar side is the *hypothenar eminence* The palm is marked by four creases two longitudinal and two transverse caused by flexions of the digits Fortunetellers lay more stress on their knowledge of the importance of these creases than they deserve

The fingers perform many complicated movements The movements of the hand and fingers are accomplished by the action of the long flexors and extensor muscles of the fingers and the flexor and extensor muscles of the carpus which arise in the forearm Perfection of motion is added by the actions of numerous short muscles

Abductor Pollicis Longus The patient tries to abduct the thumb in a plane at right angles to the palm The tendon can be seen and felt (Fig 6 32) (Radial nerve supply C C_8)

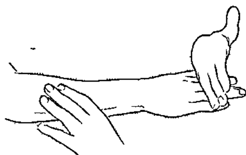


Fig 6 30 Extensor digitorum muscles Forearm and hand resting on flat surface with the palm downward Attempts are made to flex the fingers against resistance Nerve supply C_7 C_8 (radial nerve)

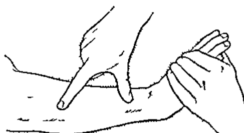


Fig 6 31 Extensor carpi ulnaris muscle Flexor surface of the arm down on flat surface Attempts are made to extend the wrist joint to the ulnar side against resistance Nerve supply C_7 C_8 (radial nerve)

Extensor Pollicis Brevis The patient resists an attempt to flex the thumb at the metacarpophalangeal joint The tendon can be seen and felt (Fig 6 33) (Radial nerve supply C C_8)

Extensor Pollicis Longus The patient resists an attempt to flex the thumb at the interphalangeal joint The tendon can be seen and felt (Fig 6 34) (Radial nerve supply C C_8)

Pronator Teres The patient extends the arm by the side and resists the examiner s attempt to supinate the hand The muscle belly can be felt and sometimes seen (Fig 6 35) (Median nerve supply C_6 C_7)



Fig 6 32 Abductor pollicis longus muscle Ulnar surface of arm flat on table Attempts are made to abduct the thumb in a plane at right angles to the palm The tendon can be felt and seen Nerve supply C C (radial nerve)

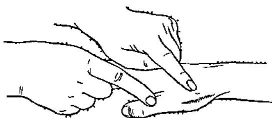


Fig 6 33 Extensor pollicis brevis muscle Palmar surface of hand on flat surface Attempts to flex the thumb at the metacarpophalangeal joint are resisted The tendon can be seen and felt Nerve supply C C_8 (radial nerve)

Flexor Carpi Radialis The patient tries to flex the wrist toward the radial side against resistance The muscle belly can be felt and the tendon can be seen (Fig 6 36) (Median nerve supply C_6 C C_8)

Flexor Digitorum Sublimis The patient resists an attempt to straighten any finger at first interphalangeal joint while the proximal phalanx is fixed The con

Supinator Brevis The patient extends the arm by the side and resists the examiner s attempt to pronate the hand (Fig 6 29) (Radial nerve supply C_5 C_6)

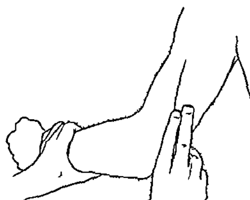


Fig 6 26 Triceps muscle The long heads or a cross section of the whole muscle can be palpated here when the patient tries to extend the arm against resistance (nerve supply C_7 C_8)

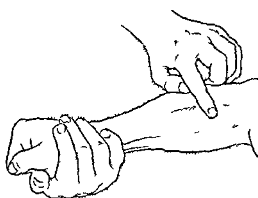


Fig 6 27 Brachioradialis muscle Arm flexed at the elbow while patient stands or sits The forearm midway between pronation and supination Nerve supply C_5 C_6 (radial nerve)

Extensor Digitorum Communis The patient resists an attempt to flex the fingers at the metacarpophalangeal joints The muscle belly can be felt and sometimes seen (Fig 6 30) (Radial nerve supply C_7 C_8)

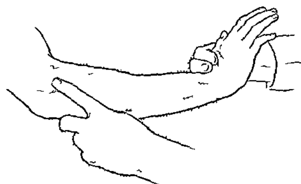


Fig 6 28 Extensor carpi radialis longus Flexor surface of arm downward on flat surface Attempts are made to extend the wrist to the radial side against resistance The muscle can be felt Nerve supply C_6 C (radial nerve)

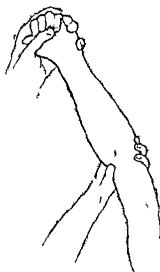


Fig 6 29 Supinator muscle Standing lying or sitting with arm extended by the side Attempts are made to prevent the hand from being forcibly pronated Nerve supply C_5 C_6 (radial nerve)

Extensor Carpi Ulnaris The patient tries to extend the wrist joint to the ulnar side against resistance The muscle belly can be felt and the tendon can be seen and felt (Fig 6 31) (Radial nerve supply C C_8)

Flexor Pollicis Longus The patient resists an attempt to extend the terminal phalanx of the thumb while the proximal phalanx is fixed (Fig 6-39) (Median nerve supply C_8 T_1)

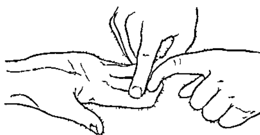


Fig 6 38 Flexor digitorum profundus muscle I and II Hand on firm surface palm upward the second phalanx is held in place and attempts are made to extend the terminal phalanx of the index finger against resistance while the middle finger is fixed Nerve supply C_8 T_1 (median nerve)



Fig 6 39 Flexor pollicis longus muscle Hand on firm surface palm up ward the proximal phalanx is held by the examiner Attempts are made to ex tend the terminal phalanx of the thumb against resistance while the proximal phalanx is fixed Nerve supply C_8 T_1 (median nerve)

Abductor Pollicis Brevis The patient s thumb is placed so that the nail is in a plane at right angles to the palm of the hand while supporting an object such as a pencil between the thumb and palm The patient then tries against resistance to bring the edge of the thumb to a point vertically above its original position The thumb nail maintains its position in a plane at right angles to the palm (Fig 6 40) (Median nerve supply C_8 T_1)

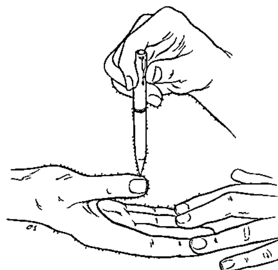


Fig 6 40 Abductor pollicis brevis muscle Hand on flat surface palm upward end of thumb over proximal palmar surface of index finger Attempts are made against resistance to bring the thumb to a position vertically above but proximal to the start ing point while the thumbnail maintains its position in a plane at right angles to the palm Nerve supply C_8 T_1 (median nerve)

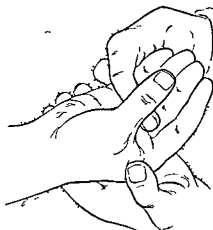


Fig 6 41 Opponens pollicis muscle The patient s arm at the elbow rests on firm surface with palm upward Attempts are made against resistance to touch the tip of the little finger with the thumb while the thumbnail remains parallel to plane of the palm The contraction of muscle is manifested by pressure exerted on examiner s right index finger Nerve supply C_8 T_1 (median nerve)

vincing sign is the ability to flex the proximal joint while the terminal joint remains flaccid (Fig 6 37) (Median nerve supply C C_8 T_1)



Fig 6 34 Extensor pollicis longus muscle Palmar surface of hand on flat firm surface Attempts to flex the thumb at the interphalangeal joint are resisted (Tendon can be seen and felt) Nerve supply C C_8 (radial nerve)

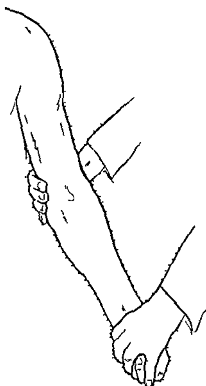


Fig 6 35 Pronator teres muscle Standing sitting or lying with arm extended by side Attempts to supinate the hand are resisted The muscle can be felt Nerve supply C_6 C_7 (median nerve)

Flexor Digitorum Profundus I and II The patient resists an attempt to extend the terminal phalanx of the index finger while the second phalanx is fixed (Fig 6 38) (Median nerve supply C_8 T_1)

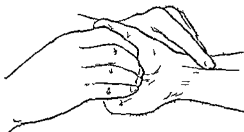


Fig 6 36 Flexor carpi radialis muscle Attempts are made to flex the wrist toward the radial side The muscle can be felt and seen Nerve supply C_6 C C_8 (median nerve)



Fig 6 37 Flexor digitorum sublimis muscle Hand on firm surface with palm upward the proximal phalanx is fixed by pressure from the examiner s fingers Attempts are made to straighten any finger at its first interphalangeal joint The proximal joint can be flexed while the distal joint remains flaccid Nerve supply C C_8 T_1 (median nerve)

flexor carpi ulnaris is intact this movement is observed even when abductor digiti minimi is paralyzed (Ulnar nerve supply C C_8)

Flexor Digitorum Profundus III and IV The patient resists an attempt to extend each finger in turn at the distal interphalangeal joint while the middle phalanx is fixed (Fig 6-45) (Ulnar nerve supply C_8-T_1)

Abductor Digiti Minimi With the back of the hand and the fingers flat upon a table the patient tries against resistance to abduct the little finger The muscle belly can be felt (Fig 6-46) (Ulnar nerve supply C_8 T_1)

First Dorsal Interosseous Muscle With the fingers and palm flat upon a table the patient tries against resistance to abduct the index finger The muscle belly can be felt and usually seen (Fig 6 47) (Ulnar nerve supply C_8 T_1)

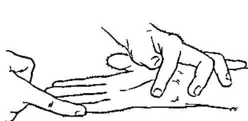


Fig 6 46 Abductor digiti minimi muscle Hand on firm surface palm upward and held in place by the examiner Attempts are made against resistance to abduct the little finger The muscle can be seen and felt Nerve supply C_8 T_1 (ulnar nerve)

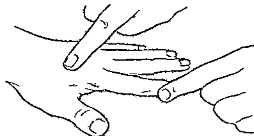


Fig 6 47 First dorsal interosseous muscle Hand on firm surface palm downward and snugly applied Attempts are made to abduct index finger against resistance Nerve supply C_8 T_1 (ulnar nerve)

First Palmar Interosseous Muscle With the fingers and palm flat upon a table the patient tries against resistance to bring the abducted index finger toward the median line (Fig 6 48) (Ulnar nerve supply C T_1)

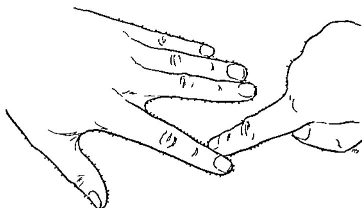


Fig 6 48 First palmar interosseous muscle Hand on firm surface palm downward and snugly applied to the surface Attempts are made against resistance to bring the abducted index finger back in place Nerve supply C_8 T_1 (ulnar nerve)

Adductor Pollicis With the thumb placed along the palmar aspect of the index finger the patient tries against resistance to retain a piece of paper between the thumb and the palm (Fig 6 49) (Ulnar nerve supply C_8 T_1)

Opponens Pollicis The patient tries against resistance to touch the tip of the little finger with the thumb while the thumb nail remains in a plane parallel to the palm (Fig 6 41) (Median nerve supply, C_8 T_1)



Fig 6 42 Lumbrical interosseous muscle of index finger Arm with palmar surface downward on firm base hand held by examiner while the first metacarpophalangeal joint is hyperextended and fixed *Attempts are made to extend the first* interphalangeal joint against resistance The muscle can be seen and felt Nerve supply C_8 T_1 (both median and ulnar nerves)



Fig 6 43 Flexor carpi ulnaris muscle Hand with fingers extended palmar surface upward on flat firm surface Examiner's hand in place indicating extent of muscle Nerve supply C C_8 (ulnar nerve)

Lumbrical Interosseous Muscle (Index Finger) With the metacarpophalangeal joint hyperextended and fixed the patient tries to extend the first interphalangeal joint against resistance The belly of the first dorsal interosseous muscle can be seen and felt (Fig 6 42) (Median and ulnar nerves C_8 T_1)

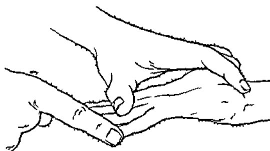


Fig 6 44 Flexor carpi ulnaris muscle (little finger) Hand resting and fixed on firm surface palmar surface upward fingers extended Attempts are made to abduct the little finger The tendon can be seen and felt Nerve supply C C_8 *(ulnar nerve)*

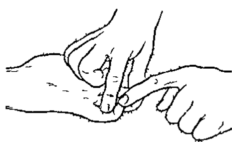


Fig 6 45 Flexor digitorum profundus III and IV of fingers Both fingers are tested as illustrated for little finger While the middle phalanx is fixed on a firm flat surface attempts are made against resistance to extend the finger at the distal interphalangeal joint while its middle phalanx is fixed The strength of the flexors can be determined by the examiner's fingers Nerve supply C_8 T_1 (ulnar nerve)

Flexor Carpi Ulnaris The patient lays the hand palm upward on the table the fingers being extended (Fig 6-43) The patient then tries to abduct the little finger fully (Fig 6 44) The tendon of flexor carpi ulnaris can be seen and felt as the muscle comes into action to fix the point of origin of abducto

reddened The uniformly reddened skin is the characteristic appearance In mild cases only the eminences of the palms and fingers are affected The color blanches on pressure with a more rapid return of the color than in the normal hand Fading of the color does not take place on elevation Definite flushing synchronous with pulse is observed when the color is partially obliterated with a glass slide

On direct microscopic examination the capillaries are said to be more numerous and dilated than normally Palmar erythema is often associated with vascular spiders

Handedness and Its Associations Handedness may be defined as the preferential use of either hand Most persons are right handed A few are left handed and some have preferences for use of the hands which are different for different tasks

There seem to be varying degrees of left handedness but only an extremely left handed person differs so greatly in motor activity from right handed persons in a right handed environment that he is set apart and is immediately recognized as such The left handed person is compelled to adapt and to adjust himself to a right handed environment In most instances the process of adjustment and adaptation is so successful that not even the affected person is conscious of a left sided tendency Consequently special tests must be used in order to recognize this peculiarity with certainty Card shuffling is a very good test for evidences of handedness

Generally total left handedness must be regarded as exceptional It is observed however that this small percentage of left handed or exceptional persons have more than their share of speech disturbances (see Stuttering Chapter 15) epilepsy and moral and physical defects

There are differences in the right and left sides of the body incident to or associated with handedness The right and the left halves of the face differ from each other and present varying degrees of asymmetry Wolff concluded that the right half of the face bears the individual expression which the person exhibits in daily life it presents those character traits and features which the person desires to portray In contrast to this the left half of the face bears the person s extra expression representing the hidden character traits which arise in the subconscious The right half of the face in right handed persons is directed by the left hemisphere which represents the seat of all rational conscious processes The right cerebral hemisphere about the importance of which very little is known controls the left half of the face the half which bears the expression of the subconscious

In the absence of any means of orientation both men and animals move in circular paths This trait of circumambulation is termed the turning tendency The turning tendency in humans is associated with handedness Right handed persons turn *chiefly toward the right and left handed ones to the left This trait* becomes apparent when a right handed and a left handed person are together and are lost in the woods The right hander wishes to go to the right and the left hander to the left and thus an argument ensues

The natural turning tendency in either man or animal is lost when the individual is wounded The path after the person or animal is wounded—shot—is straight away from the scene of the tragedy

Regarding the postural and motor acts which are peculiar to man and which consist in interaction of two extremities some individuals will favor one side and others will reveal no preference for either side and thus these acts cannot be associated with handedness However the manner in which a person folds the hands and crosses the arms rather specifically indicates handedness For example the superposition of the left thumb upon the right when the hands are folded is an indication of left handedness

The foot which is passed forward to test the strength of ice or the softness of the ground before treading on it is an indication of footedness It may be observed that the footedness is contralateral to the handedness For instance a right handed

Opponens Digiti Minimi Fingers extended the patient tries to make a cup of the palm and to carry the fifth finger in front of the others (Fig 6 50) (Ulnar nerve supply C_8 T_1) *



Fig 6 49 Abductor pollicis muscle Hand on firm surface palm upward fingers extended thumbnail in a plane at right angles to the palm Ability to hold a piece of paper against resistance indicates intact muscle Nerve supply C_8 T_1 (ulnar nerve)

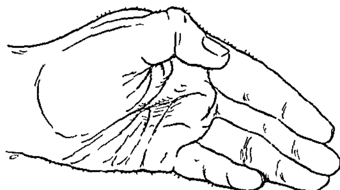


Fig 6 50 Opponens digiti minimi muscle The ulnar aspect of the arm rests on flat surface with thumb upward Attempts are made to form a cup of the palm while carrying the little finger in front of the others Nerve supply C_8 T_1 (ulnar nerve)

The Palmar Fascia The palmar fascia or aponeurosis is the deep fascia of the hand It consists of a thick triangular middle portion and two thin lateral portions which cover the thenar and hypothenar eminences This fascia is often involved in injuries and infections of the hand Dupuytren s contracture is a congenital anomaly involving the fascia of the hand (see Disease of Fascia p 273)

Palmar Erythema The palmar surface of the hand is less pigmented than is the dorsal surface In the absence of callus the lividity of the palmar and dorsal surfaces of the hand is equal A morbid redness of the skin of the palmar surface is palmar erythema a condition due to dilatation of the capillaries in the pads of the palm and fingers The covering skin is normal

Palmar erythema on rare occasions may be present in an otherwise normal person It is usually associated with some sort of chronic illness and often an illness in which there is an associated hypoalbuminemia It may be present in liver disease and during the last trimester of pregnancy However it may be associated with chronic diseases of many kinds

Palmar erythema occurs in both men and women There are no symptoms However the patient may be concerned about the appearance of the hands

Examination reveals symmetrically reddened eminences of the pads of the terminal digits the palms and sometimes the soles of the feet The transverse creases of the fingers may be of a deeper color than the adjacent skin The redness may extend over the finger tips to the nail beds and laterally over the distal phalanges The color may be a diffusely mottled redness small telangiectatic spots or uniformly

* Acknowledgment is made to the Controller of Her Britannic Majesty s Stationery Office for permission to make line drawings of photographs of muscles in action in War Memorandum No 7 reprinted 1945

Of the *chronic inflammations* in and about the *wrist* chronic infectious arthritis is very common If one wrist alone is affected and no other joints in the body are involved tuberculosis of the joint is probable It is easily distinguishable from tuberculous tenosynovitis since in the latter the swelling is only on one side of the wrist usually the volar side and is most marked not at the level of the wrist itself but proximal and distal therefrom The fingers are slightly flexed and there is no volar subluxation of the hand as in tuberculous arthritis

Of the chronic inflammations of the *hand* and *fingers* the common ones are (1) tuberculosis of the tendon sheaths especially of the flexor tendons (2) gonorrheal tenosynovitis (3) the stenosing tenosynovitis (radiating pain in the thumb and forearm on use) due to stenosis of the compartment of the tendon sheath for the extensor hallucis brevis muscle and the abductor hallucis longus muscle over the styloid process of the radius and (4) the spindle shaped swellings of the metacarpal bones or phalanges due to tuberculosis or to syphilis

Tenosynovitis (*Perimyotendinitis*) Traumatic tenosynovitis is distinct from pyogenic or granulomatous involvement of the tendon sheaths The term tenosynovitis applied to the usual crepitating condition found proximal to the wrist joint is a misnomer The actual site of the lesion is well above the upper limit of the tendon sheaths at the wrist

The local manifestations suggest that the lesion is essentially a perimyotendinitis involving the more proximal musculotendinous junction in the forearm It affects mostly factory workers agricultural workers and housewives The tendons most frequently affected are the radial extensors of the wrist (extensor carpi radialis longus and brevis) and the abductor pollicis longus or extensor pollicis brevis either alone or in combination The main factors in the etiology are unaccustomed movement resumption of work after absence repetitive stereotyped movement and local strain The prognosis is good and cessation of work is neither desirable nor necessary

Neoplasms The wrists are rarely affected by neoplasms The commonest tumors of the wrist are the osteophytic outgrowths from the bases of the distal phalanges in certain elderly persons The wrist joint may be affected by sarcoid and giant cell tumors

Diseases Due to Trauma or Physical Agent ***Abscesses of the Hand and Fingers*** Purulent collections in the palm of the hand either are situated beneath the palmar fascia or are connected with the sheaths of the flexor tendons

When pus collects beneath the palmar fascia it is limited in the directions in which it can spread Pus originating beneath the thick middle triangular portion of the fascia may point in one or more of several places (1) on the inner side at the hypothenar eminence (2) in the web of the thumb (3) on the anterior surface of the forearm above the wrist and (4) on the webs of the fingers Also in rare instances (5) it may burrow between the distal extremities of the metacarpal bones and show on the back of the hand and (6) if there should be as is sometimes the case tiny gaps in the fascia a small amount of pus may accumulate above the palmar fascia and between it and the skin so that there is a collection of pus both above and below the fascia

If the sheaths of the tendons of the hand or fingers become infected the pus passes along the tendon as far as the sheath extends Since the extent of the tendon sheaths varies it is well to recall that when suppuration occurs in the sheath of the thumb or little finger it is much more serious than in the other three because the pus tends to pass directly upward involve the palm and extend to the arm When suppuration involves the index middle or ring finger it stops when it reaches the vicinity of the metacarpophalangeal joints and involves the palm only It may penetrate the carpal bursa

When the end phalanx becomes infected the affection is known as a *felon* The

person may twist and prefer use of the left foot for purposes of exploration of the safety of the terrain for walking but will kick a football with the right foot

Abnormalities of the Hand The *spade hand* large coarse thick fingered with broad nails is seen as an evidence of myxedema in which the enlargement affects mainly the soft parts and in acromegaly in which the enlargement affects the bones

The *clawhand* also called *main en griffe* (griffin hand) is a deformity which occurs in consequence of paralysis and atrophy of the interossei and lumbricales muscles Paralysis of these muscles leads to a dorsal extension of the proximal phalanges with flexion of the others and when atrophy takes place the clawhand results This abnormality suggests the existence of a neuritis of the median and ulnar nerves particularly the latter or progressive muscular atrophy Similar deformities may be due to amyotrophic lateral sclerosis (the spastic form of progressive muscular atrophy) syringomyelia and very rarely the adult type of chronic anterior poliomyelitis

Atrophy of the muscles of the hand and forearm occurs in brachial palsies but also to a slighter degree in cerebral paralysis in consequence of disuse and lower motor neuron lesions

Coldness of the hands and feet with or without a tendency to sweating if persisting for weeks or months is most commonly due to neurasthenic conditions Sudden or transient coldness of the extremities is observed in many persons especially those of a nervous temperament under excitement or anxiety as well as in shock collapse hemorrhages and premonitory chilliness of rising fever Coldness of the hands is also observed in Raynaud s disease and Buerger s disease It is necessary to discriminate between actual lowering of the temperature palpable to the observer and a subjective sensation of coldness The latter is a paresthesia

Excessive *sweating of the hands* may be due to nervous tension toxic goiter and occasionally progressive muscular atrophy

In determining the existence of enlarged joints it must not be forgotten that atrophy or wasting of the soft parts may cause an apparent increase in the size of the joints

Diseases of the Wrist and Hand The more distinctly medical diseases of the wrist and hand include the inflammations the neoplasms and trauma

Inflammations of the Wrist and Carpal Region The *acute inflammations* of the wrist joint and its neighborhood (chirarthritis) are (1) acute arthritis of the wrist due to acute rheumatic fever or other forms of acute infectious arthritis and (2) acute inflammation of the vaginae mucosae (tenosynovitis) secondary to a penetrating wound of a finger to a bite (human or animal) or other injury or sometimes to metastatic infection

In tenosynovitis it is chiefly the movement of the fingers that is disturbed whereas in arthritis of the wrist it is chiefly movements of the hand as a whole that are concerned moreover in arthritis there is everywhere about the affected joint tenderness on pressure while in tenosynovitis only the side affected is tender Again traction or compression in the long axis of the extremity is painful in arthritis not in tenosynovitis Extension of the inflammatory process in the longitudinal direction is characteristic of tenosynovitis in arthritis the inflammation remains limited to the joint region Sometimes of course arthritis and tenosynovitis occur together

A painful swelling in the region of the long extensor of the thumb accompanied by crackling on palpation over the tendon and muscle is the so called tenosynovitis crepitans, it is a chronic inflammation of the vaginae mucosae of the tendon and of the surrounding tissues

The metacarpal and phalangeal joints are often the site of arthritis The proximal phalangeal joints especially undergo spindle shaped thickening in chronic infectious arthritis (rheumatoid)

Eczema of the Hands Eczema of the hands may be self limited as in occupational eczema of the hands in which the skin becomes hardened or it may be a permanent state in which remissions and exacerbations alternate A familial history of eczema or a history of infantile eczema is significant because it indicates an allergic state

There are many known causes for eczema of the hands and probably just as many unknown causes The known causes are fungous infections and their reactions occupational irritants dietary factors neurodermatitis dyshidrosis (pompholyx) and drug eruptions The presence of an acute vesicular eruption indicates that the skin is hypersensitized and is prone to react unfavorably to therapeutic agents that are not mild and soothing

Eczema or dermatitis of the hands consists of a vesicular or scaling oozing or weeping crusted or infected eruption of the hands In the acute phase it is characterized primarily by vesicles and erythema when subacute by scaling and erythema and in the chronic phase by thickening or lichenification and scaling

Diseases of the Nails ***Congenital Abnormalities*** Congenital absence of the nails (anonychia) is very rare It may be associated with congenital ichthyosis if so nails may or may not appear later in life

The presence of extremely small otherwise apparently normal nails is termed micronychia Extremely large nails (macronychia) appear as a congenital abnormality they also develop later in life in the course of acromegaly or macrodactylia and may at times be associated with clubbing of the fingers

Pachyonychia is a congenitally thickened and discolored nail which may also be abnormally hard and longitudinally striated

Nails may be present in abnormal situations for instance on the palmar surface of a finger or the plantar surface of a toe This condition is known as onychoheterotopia

Infections Tuberculosis anthrax leprosy virus of verruca vulgaris syphilis and mites of scabies may cause nail changes Most cases of suppurative paronychia however are caused by staphylococci whereas nonsuppurative paronychia often results from moniliasis

Syphilis affects the nails in a variety of ways However these changes are not constant or definite The diagnosis of syphilis is made on the presence of positive serologic reactions which are associated with other evidence of syphilis elsewhere in the body If all of these findings are present and there is no evidence for any other causative disease the changes in the nails may be attributed to syphilis

Fungous disease or ringworm of the nails—onychomycosis—causes severe disturbances of nail growth and local destruction of the nails of the toes or fingers Nail foci may provide sources of infection or reinfection to the glabrous skin The abnormalities of nail structure may be so severe as to be incapacitating as in mycotic onychauxis and onychogryposis

The fungi causing the different types of onychomycosis are many and varied Trichophyton Epidermophyton *Candida albicans* (*monilia*) Penicillium Aspergillus (see Diseases of the Skin Chapter 8)

When a nonsuppurative paronychia is present moniliasis caused by *Candida albicans* is to be suspected especially in fruit pickers and handlers dishwashers housewives dentists and others whose nails are wet during prolonged periods

Because of the close resemblance of onychomycosis to psoriasis the diagnosis always depends on microscopic and cultural identification of the fungi Positive cultures are obtained from 1 of every 5 who have the disease

Trauma Schwartz Tulipan and Peck have considered in detail the effects of trauma to the nails Hemorrhage beneath the nail plate (subungual hematoma) may be caused by a bruise and the nail may subsequently be loosened and fall away Deep splinters may cause longitudinal white streaks a form of leukonychia Spoon shaped nails (without regrowth) may form or in elderly patients such a nail may

skin over the pad of the finger sends off fibrous bands or fibrils which are attached to the parts beneath An infection of the skin can pass through it and involve the fatty tissue beneath The pus is often unable to break through the skin Since the fibrous bands prevent swelling it burrows and involves the periosteum and thence to the region of the joint where it may enter the sheath Once the pus is within the tendon sheath it extends as far as the sheath goes

In pursuit of his labors man is exposed to various hazards that deform his body and cripple his hands The changes which affect the hand under the influence of mechanical chemical thermal electric and atmospheric stimuli are numerous These changes may be characteristic of certain trades so that they may be regarded as trade stigmas For instance hands which are in constant contact with water may be affected by an alteration of the skin characterized by softening chapping and often ulceration with veritable destruction of the parts

The trade stigmas of the hands have been classified by various outlines Oppenheim long ago enumerated these stigmas as stains or coloring A more recent classification has been enumerated by Schwartz Tulipan and Peck as stratifications excoriations telangiectases nail changes pigmentation callosities bursae cicatrices and tattooing

The nails frequently exhibit trade stigmas The right thumbnail in women who remove the indigo fruit from its shell is long In watchmakers and clockmakers this nail is short The nails of laundresses and lens grinders are short and cracked

The hands become covered with particles of the material used at the place of work Thus deposits of coal dust forming indelible blue black scars enable one to recognize the coal miner who when working soils every scratch with coal dust *Similar deposits in the skin are found among metal workers millers and stone masons*

Superficial wounds excoriations and rhagades are to be found among seamstresses dressmakers cobblers and glassworkers The hands of glassblowers commonly exhibit wheals flat plaques rhagades and warts (For further discussion of trade stigmas and mutilating affections of the hands see Chapter 24)

Ankylosis of the Hand The finger and wrist flexors are stronger than the extensors The adductors of the thumb are stronger than the abductors *The wrist or* fingers stiffen in a flexed position or the thumb in an adduction deformity when prevented from use

Prolonged immobilization is probably the most frequent cause of stiffness in the hand because of the necessity of continuous splinting in injuries of the arm or the hand In nerve injuries in loss of substance and in suppurations such as palmar abscess the usefulness of the hand is sometimes totally lost Without the use of the hand grasp all the wide excursions of the elbow and shoulder are useless Impediment in the shoulder or the elbow also limits use of the hand

A *snapping finger* is produced by some obstruction to the action of the long dorsiflexor tendons under the ligament across the joint If painful or disabling determination of the nature of the obstruction is made

Ankylosis or Loss of the Thumb Without use of the thumb the hand is left with only parallel digits and the palm is the only opposing force in the act of gripping

The fingers bear the evidence of the influence of the cerebral cortex on the acquisition of willed movements They possess the tactile senses so necessary for the most technical accomplishments of the hand or the coarser squeezing duties which require strength and action Without the fingers or thumb the hand becomes useless Without individual action the finer functions of the fingers are lost There fore the interruption of perfection in one digit disturbs the synchronization and proficiency of the entire hand

Nutritional Deficiency Egg shell nail is commonly seen in avitaminosis A Koilonychia (spoon nails) may be present in association with dysphagia glossitis and hypochromic anemia (Plummer Vinson syndrome) an iron deficiency disease Beau s lines and thinning and brittleness of the nails occur in many different types of malnutrition and in severe systemic disorders

Changes in Dermatoses LICHEN PILARIS SEU SPINULOSUS In lichen pilaris (keratosis pilaris) atypical nail changes may occur

KERATOSIS FOLLICULARIS (DARIER S DISEASE) The nails may become friable and split and onychauxis onychogryposis paronychia and longitudinal ridges may develop

EXFOLIATIVE DERMATITIS Onychomadesis is of frequent occurrence in exfoliative dermatitis as in drug sensitivity However regrowth often takes place

ALOPECIA AREATA In alopecia areata interruption of nail growth producing Beau s lines thinning and increased friability may be present Regrowth almost invariably occurs

ECZEMA Nail changes in eczema are highly varied ranging from minor disturbances of growth and distortion of structure to almost complete erosion and onychomadesis Suppurative paronychia is frequently associated with eczema

Nail changes occasionally occur in psoriasis

Tumors Involving the Nails Nails can be deformed in varying degrees by verruca clavus and exostoses of the phalanges A chondroma in the terminal phalanx causes deformation of the nail A glomus tumor may arise immediately beneath the nail bed on which it may encroach to become visible as a bluish discoloration under the nail plate A glomus tumor is exquisitely painful A simple angioma may be observable as a red discoloration in the nail bed or about the nail Occasionally a melanoma originates from the matrix or nail fold A fibroma may be observed beneath the nail bed or about the nail

DISEASES AFFECTING THE ARM AND HAND AS A WHOLE

Arachnodactyly The individual afflicted with arachnodactyly according to Parker and Hare is tall with excessively long arms and legs long thin spider like hands meager subcutaneous fat under developed atonic musculature and often ectopia lentis The emaciation accentuates the bony landmarks and these in turn serve to emphasize the disproportionate length of the extremities as compared to the trunk The delicately elongated spider like fingers give this syndrome its name The feet and toes are likewise long and slender Deformities of the joints are common Pes planus contractures hammer toes webbing and sometimes abnormally mobile patellas occur The frequently associated scoliosis kyphosis winged scapula and deformities of the sternum are considered to be the result of the laxity of the ligaments and atonic musculature The skull of the arachnodactylic patient tends to be dolichocephalic with prominent supra orbital ridges frontal bossing a pointed chin and either a prominent or a broad somewhat flattened nose The face is usually drawn wrinkled and old looking often with a melancholy expression The ears are frequently enlarged Approximately 50 per cent of the patients have a partial dislocation of the lens usually upward Congenital heart anomalies frequently accompany the disease and consist mainly of a patent foramen ovale and other interauricular septal defects In the infant or rapidly growing child this syndrome may resemble rickets The abnormally long and slender extremities the absence of flared epiphyses normal blood calcium and phosphorus and perhaps obscure anomalies in the parents should lead one to suspect arachnodactyly The symptoms of weakness fatigue moderate dyspnea on exertion malnutrition and a coarse systolic murmur in early childhood may focus attention on the cardiovascular system to the exclusion of what appear to be lesser anomalies This syndrome is not considered to be of endocrine origin

Venous Obstruction in Upper Extremity An arm of an otherwise healthy person may become diffusely swollen and discolored It has been suggested that

become wrinkled and hyperkeratosis of the periungual tissues may form and malignant degeneration with the formation of epithelioma may ensue

Chemical Agents There are a number of chemical agents among which are included soap nail polish polish remover nail polish undercoats dyes and industrial chemicals which injure the nails Pigmentation or discoloration of the nail may be produced by contact with *topical medicaments* such as gentian violet mercurochrome silver nitrate chrysarobin silver containing compounds arsenic tobacco and many chemicals used in industry However in industry the offending substances are known and the hands are usually protected

Soaps nail polish bases (undercoat) fabric fibers (wool silk) and chemicals cause allergic lesions after repeated contact The lesions vary from leukonychia and minor ridging to marked destruction of the nail plate and inflammation of periungual and subungual tissues Often these lesions resemble those of mycotic infections and psoriasis

Nail discoloration and dystrophy may be due to nail polish and nail polish bases or undercoats Sulzberger and others have observed the clinical features associated with allergic eczematous reactions of the nail bed due to synthetic resin rubber undercoats and they have enumerated the following features (1) a discoloration diffuse or punctate of the distal portion of the nail which ranges from yellow through blue to black (2) separation of the distal portion of the nail from its bed (onycholysis) (3) subungual hyperkeratosis (4) erythema edema and scaling of the fingertips under the free edge of the nail and subjective symptoms in the areas may occur if the nail which has an undercoat is subjected to repeated trauma

Nail discolorations occur as the result of injury to the tips of the fingers The nails become separated from their beds—a condition known as onycholysis Nails may become discolored and may separate if repeatedly subjected to external heat A splinter of wood glass or metal often tears the nail away from its bed and causes discoloration and perhaps separation

Trauma and Physical Agents Workers who use power driven drills or hammers are subject to development of hyperkeratoses beneath the nails Gardeners laborers textile workers and those who handle wool often suffer from nail infections following trauma (Schwartz Tulipan and Peck have considered in detail diseases of the nails in industry)

The nail matrix is highly *radiosensitive* Nail changes may be the first observable signs of overexposure to roentgen rays as well as to the emanations from radioactive elements such as radium uranium plutonium and many radioactive isotopes such as those of phosphorus and iodine

Enumeration of the early disturbances of nail growth caused by irradiation according to Schwartz are (1) thinning softening and increased friability of the nail (2) longitudinal splitting (3) discoloration and (4) varying degrees of atrophy As time passes after irradiation transverse ridging (Beau s lines) and loss of the nail occur

Vascular Disease In both vasospastic and obliterative vascular disease the nails undergo specific changes indicative of the lack of adequate circulation

In vasospastic conditions according to Allen a pterygium develops (1) the nail fold becomes thinner and the cuticle widens (2) the normally distinct boundary between nail fold and cuticle as well as that between cuticle and nail plate is lost

Pterygium is a common condition of the last two toe nails of apparently normal individuals It is also common however in Raynaud s disease and scleroderma

In thromboangiitis obliterans severe ischemia causes distortion of the nail plate The linear growth is slowed or imperceptible and the nail plate becomes thick rough dark and distorted Changes in the nails occur as the result of peripheral nerve lesions as in leprosy

Endocrine Disorders In hypothyroidism the nails become brittle growth is slowed and onycholysis occurs In hyperthyroidism atrophy of the nails is common together with onycholysis and longitudinal ridging Koilonychia may be prese[illegible] roidism Onychogryposis (claw nails) may occur in acromegaly

limb against resistance The muscle bellies can be felt and sometimes seen (Fig 6 56B) * (Superior gluteal nerve supply L_4 L_5 S_1)

The Hip Joint and Monarticular Disorders The hip joint is a ball and socket joint The functions of the hip are mobility and support For the necessary support and limitation of extent of movements of the hip there are strong bandlike ligaments uniting

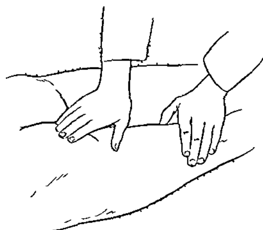


Fig 6 54 Adductor muscles of leg Lying on back with leg extended and abducted patient attempts to adduct the leg again t resistance The muscles can be seen and felt Nerve supply L L_3 L_4 (obturator nerve)

the bones In the upright position the center of the weight of the body falls in front of the axis of rotation of the hip joint When a person falls it is usually forward Falls in any other direction are indicative of complete loss of balance

The head of the femur is 2 inches (about 5 cm) in diameter and spherical in shape The acetabulum is deep The joint is airtight and the femur is held in place by suction and ligaments

Fig 6 55 Gluteus maximus muscle Lying with face downward abdomen and leg flat on firm surface patient attempts against resistance to lift knee upward The muscle can be seen and felt Nerve supply L_4 L S_1 S (inferior gluteal nerve)



The great pressure to which the articulating surfaces of the hip joint are subjected requires special lubrication of these surfaces and this is supposed to be furnished by the ligamentum teres and haversian glands

Congenital Malformations of the Hip In the anamnesis a history of limping or of waddling from earliest childhood can be obtained

DISLOCATIONS Congenital luxations or dislocations of the hip are most frequently posterior in position and are characterized by excessive mobility of the thigh The manifestations of this deformity are no eversion no flexion on lying down and usually a high grade of lumbar lordosis on standing The acetabulum may be shallow The head of the femur is deformed and it occasionally is superior but

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Sartorius The patient lies on the back with the hip laterally rotated and tries to flex the knee against resistance The muscle belly can be felt and sometimes seen (Fig 6 52) (Femoral nerve supply L L_3)

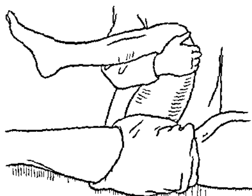


Fig 6 51 Iliopsoas muscle Lying on back with the knee flexed while the leg is supported at less than a right angle between the thigh and trunk Patient attempts to flex the hip against resistance Nerve supply L_1 L L_3 (femoral nerve)

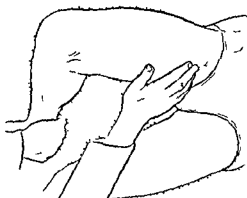


Fig 6 52 Sartorius muscle Lying on back with leg to be tested laterally rotated at the hip patient attempts to flex the knee against resistance Nerve supply L L_3 (femoral nerve)

Quadriceps Femoris The patient tries to extend the knee against resistance The muscle bellies can be felt and often seen (Fig 6 53) (Femoral nerve supply L L_3 L_4)

Adductors The patient lies on the back with the knee extended and tries to adduct the leg against resistance The muscle bellies can be felt (Fig 6 54) (Obturator nerve supply L L_3 L_4)

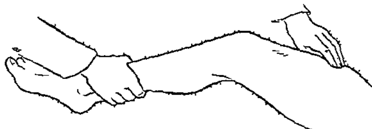


Fig 6 53 Quadriceps femoris muscle Lying on back with knee flexed patient attempts to extend the knee against resistance The muscle can be seen and felt Nerve supply L L_3 L_4 (femoral nerve)

Gluteus Maximus In the prone position the patient tries against resistance to lift the knee The muscle belly can be felt and often seen (Fig 6 55) (*Inferior* gluteal nerve supply L_4 L_5 S_1 and S)

Gluteus Medius and Minimus and Tensor Fasciae Latae (Rotation) To test internal rotation at the hip joint the patient is in the prone position with the knee flexed to a right angle and tries to carry the foot laterally against resistance The muscle bellies can be felt and sometimes seen (Fig 6 56A) (Superior gluteal nerve supply L_4 L , S_1)

Gluteus Medius and Minimus and Tensor Fasciae Latae (Abduction) To test abduction the patient lies on the back with the leg extended and tries to abduct th

Coxa vara is to be distinguished from (1) congenital dislocation of the hip (2) beginning coxitis and (3) the effects of trauma

Infections of the Hip Joint Hip disease coxitis or coxalgia in its early stage is characterized by pain limitation of motion and limping The pain may be limited to the hip itself or referred The hip is supplied by branches of the anterior crural sciatic and obturator nerves and since these also supply the region of the knee disease of the hip often causes pain to be referred to the region around the knee joint

In an early stage the limitation of motion is due to muscular contraction The limb is held in a position of flexion abduction and slight external rotation In mild cases the limitation is present as a reduction only in the normal extent of move ments The abnormal changes are recognized by inspection measurements and comparison with the opposite healthy limb

On inspection from behind the gluteal fold is small over the affected side and the buttock is flattened The flattening of the buttock is caused by the flexion of the hip An inequality in the lower limbs whether due to shortening or to malposition such as flexion will be visible at once by an inequality of the gluteal folds one being higher than the other Flexion deformity is recognized when the patient is standing by the bending at the hip joint and by the lordosis or hollowing of the back In flexion deformity of the hip when recognized the degree can be measured while the patient is in the recumbent position by the difference in the length of the legs

The patient being flat on the back the pelvis is made level by seeing that a line joining the two anterior iliac spines is at right angles to the median line If abduction is present the limb points away from the median line It cannot be brought straight down parallel with the sound leg without tilting the pelvis If measured from the umbilicus to the internal malleolus the affected leg measures more than the sound one This is called apparent lengthening If when both limbs are placed in the same degree of abduction and are measured from the anterior iliac spine to the internal malleolus they measure the same there is no real shortening

In advanced disease adduction is commoner than abduction Adduction produces an apparent shortening as shown by measurement from the umbilicus to the internal malleolus if the sound limb is placed in the same degree of adduction as the infected one the distances from the anterior iliac spines will show no actual shortening unless there is a loss of bone or displacement at the hip joint The pelvis instead of being tilted down on the diseased side is tilted up Flexion is usually more marked and the foot is usually inverted instead of everted

In the diagnosis of the acute inflammations of the hip and its neighborhood it is necessary to distinguish acute inflammations of the hip joint itself from osteo myelitis of the shaft of the femur In the latter cautious passive movement of the hip is possible and there is local tenderness below the trochanter Also to be dis tinguished are iliac abscess femoral or inguinal lymphadenitis and acute phlebitis of the femoral vein

If acute arthritis is present it may be due to acute rheumatic fever to other kinds of infectious arthritis to syphilitic arthritis or to very acute tuberculous coxitis

CHRONIC COXITIS Of the chronic infections of the hip joint in childhood by far the most important is tuberculous coxitis the common hip joint disease of child hood In adults nontuberculous arthritis is commoner including avascular necrosis

In *tuberculous coxitis* the child begins to show a painful limp the gluteal fold disappears and the muscles of the thigh begin to atrophy Sometimes the child com plains of pain in the knee rather than in the hip but examination of the knee joint does not reveal anything abnormal there On examination of the child lying on its

usually posterior to the acetabulum It can be moved backward and forward on the pelvis The neck of the femur is twisted on its shaft

There is shortening of the leg on the affected side when measurement is made from the anterior superior iliac spine The anterior iliotrochanteric angle is diminished or lost The tip of the trochanter lies abnormally high above the Roser Nelaton line By palpation it can be recognized that the head is not in its normal position beneath the femoral artery The superior border of the trochanter is on a level with the anterior superior iliac spine Roentgenologic examination is necessary to ascertain accurately the position of the head and whether or not the bones possess their normal shape

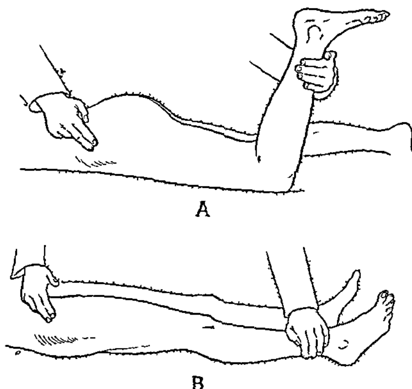


Fig 6 56 A gluteus medius and minimus and tensor fasciae latae muscles as internal rotators Lying with face downward belly flat leg flexed at knee patient attempts to carry the foot laterally against resistance The muscle can be felt Nerve supply L_4 L_5 S_1 (superior gluteal nerve)

B gluteus medius and minimus and tensor fasciae latae muscles as abductors Lying flat on back on firm surface legs extended patient attempts against resistance to abduct the leg The muscle can be felt Nerve supply L_4 L_5 S_1 (superior gluteal nerve)

The abnormal mobility distinguishes congenital dislocation of the hips from bilateral coxa vara due to rickety curvature of the femur Poliomyelitis may cause a paralytic limp resembling congenital dislocation but the roentgenologic examination reveals the head of the femur in the acetabulum

Coxa Vara Coxa vara is a curvature of the neck of the femur causing adduction of the thigh and apparent shortening of the leg The changes in the position of the neck of the femur in coxa vara may be due in childhood to rickets in later life to the carrying of heavy burdens or more rarely to osteomalacia The coxa vara of childhood is usually bilateral that of adults may be either unilateral or bilateral The signs of coxa vara are (1) anterior position and abnormally high position of the trochanter (2) lateral rotation and limitation of abduction of the thigh (3) medial rotation and flexion and (4) when unilateral shortening and limping

Roentgenograms are diagnostic and should be made unless it is unwise to move an aged infirm patient

Ankylosis of the Hip Joint Complete ankylosis of the hip joint is somewhat compensated by a flexible lumbar portion of the spinal column The unfavorable positions of ankylosis of the hip joint are considered to be severe flexion with adduction and internal rotation since these positions interfere with standing or walking because of the shortening that they produce which affects lateral equilibrium

A favorable position of ankylosis is that approaching extension in the long axis of the body In this position standing and walking are possible but sitting is hampered Ankylosis in flexion however is favorable to the sitting position

Varying degrees of ankylosis of the hip are caused by (1) Contusions or sprains which may result from a severe blow or fall with the impact over the trochanter or imparted through the shaft A single dislocation does not induce more than temporary ankylosis Repeated dislocations may cause stiffening (2) Fracture of the acetabulum or of the head or the neck of the femur may cause stiffness (3) Continuous immobilization over a long period of time may permit of atrophy adhesions and stiffness (4) Suppuration in the hip joint usually results in complete ankylosis or a disability just as bad resulting from joint pain on movement (5) Rheumatic arthritis in its many forms frequently affects the hip joint

Adduction in a leg that is fully extended at the hip produces disability of standing balancing walking jumping and running The disability in all these activities is increased by the shortening effect which increases as the adduction deformity increases

In flexion deformity abduction is an advantage because it decreases the lengthening effect unless there is external rotation

Internal rotation increases the disability in the performance of all the duties of the leg Outward rotation produces no disability so long as the toes can reach the normal vertical weight bearing line When the toes cannot reach the midline disability becomes more severe and especially when there is flexion deformity outward rotation adds to the disability Internal rotation with flexion brings the toes into a more convenient position for weight bearing (McBride)

Tumors of the Hip and Thigh Of the neoplasms of the hip and thigh those involving the muscle include angioma fibroma and sarcoma all of which must be differentiated from tuberculous myositis gummatous myositis and muscle hernias Those originating in the bone include cartilaginous exostoses enchondromas osteomas and myeloid sarcomas Such tumors of the bone must be distinguished from osteomyelitis and fractures A spontaneous pathologic fracture of the bone may occur in sarcoma

A positive diagnosis as to the nature of a tumor is established by biopsy

Diagnosis of Hip Joint Disease In hip joint disease there may be no symptoms referable to the hip The pain may be referred to the knee As the disease advances there develops a limp as the result of a shortening of the leg on the affected side As the leg shortens the pelvis tilts and a scoliosis develops

In a limp from a congenital dislocation of the hip the patient steps forcibly forward on the affected side In these limps the patient obviously is not afraid of producing pain in the leg or foot but limps in order to place himself more firmly on the affected leg If the dislocation is bilateral a waddling gait develops A limp may result from painless rigidity of one hip joint as from a healed coxitis injury or rickets the whole extremity including one half of the pelvis being moved forward as a unit Bilateral stiffness of the whole lower extremity occurs in extreme bilateral coxa vara rheumatoid arthritis and from the prolonged use of casts A limp results when there is pain in some one of the joints on the affected side on movement as a result all the joints of that side may undergo muscular fixation The patient avoids

back with the legs straight out there is obvious lordosis of the lumbar region of the spinal column If this lordosis is overcome by strong flexion of the thigh of the healthy side there will be involuntary flexion of the thigh of the affected side when tuberculous coxitis exists (Thomas test) There is fixation of the affected extremity and the pelvis these parts moving as a whole

During the progress of the disease a cold abscess may form pointing in the following order of frequency (1) in front below the anterior superior iliac spine (2) lateralward and (3) backward Tenderness on pressure can be elicited at the front of the joint below the middle of Poupart's ligament and compression from the trochanter is painful

Roentgenologic examination in the earliest stages of tuberculous coxitis may not reveal definitely positive findings except for an osteoporosis due to disuse After the disease has advanced and has involved the cartilage and bone the roentgenologic findings are characteristic and diagnostic

Tuberculous coxitis is distinguished from (1) subacute forms of simple infectious coxitis (2) chronic infectious arthritis (3) congenital dislocation of the hip (4) the different forms of coxa vara (5) caries of the spinal column and of the pelvis with cold abscess formation (6) periappendicular abscess and perinephric abscess (7) hydrops of the bursa iliaca (8) sciatica and other neuralgias and (9) hysteria (Brodie's joint)

Nontuberculous chronic coxitis may be either rheumatoid arthritis or osteoarthritis Occasionally the hip is involved in a neuropathic arthropathy such as tabes dorsalis or syringomyelia

Avascular degeneration of the head of the femur may occur from unknown causes The course is a chronic one characterized by pain which is often referred to the knee The roentgenologist can usually suggest the correct diagnosis

Injuries of the Hip Joint If after an injury to an aged person in the home roentgenologic examination is not readily available the following procedures may be helpful diagnostically The position and attitude of the injured extremity should be inspected for this inspection alone will often give a clue to the nature of the injury Inspection is followed by measurements to ascertain whether or not there is shortening and if so whether it is supratrochanteric or infratrochanteric

The position of the trochanter is determined by means of the Roser Nélaton line or by outlining Bryant's triangle The Roser Nélaton line joins the anterior superior spine of the ilium to the tuberosity of the ischium normally the greater trochanter of the femur lies in this line Bryant's triangle is constructed while the patient is lying on the back Let fall a perpendicular line from the anterior superior spine to the table The other side of the triangle is a line joining the anterior superior spine and the tip of the greater trochanter The base of the triangle is a line running horizontally from the tip of the trochanter to the perpendicular line In fracture of the femur the trochanter is elevated when the neck of the femur is fractured the base of the triangle is shortened

If it is determined that there is shortening of 1 inch (about 2 5 cm) and there is good reason to believe it was not present before the injury a dislocation or a fracture has occurred If the shortening is supratrochanteric there has been a dislocation or a fracture of the neck of the femur if it is infratrochanteric there has been a fracture below the tip of the trochanter If the trochanter is abnormally high it indicates a dislocation or a fracture of the neck If the trochanter occupies its normal position usually there is no dislocation or if there is one it must be situated below the trochanter

The active movements of the leg are examined by desiring the patient to lie flat on the back and lift the injured leg If the injured leg can be lifted there is no dislocation or fracture If the thigh can be flexed with difficulty without raising the heel from the bed an impacted fracture may have occurred in which event there will be limitation of active medial rotation

thus moved and if such an attempt is made a sharp pain ensues A cirsoid femoral vein is soft and yields to pressure and will collapse when the leg is elevated

THE KNEE JOINT

The knee joint is primarily a hinge joint and is firmly and strongly braced by ligaments Its movements are extension and flexion It can be extended to a straight line and flexed until the thigh and the upper portion of the leg come in contact Any detectable rotary movements of the knee when fully extended are associated with injuries

The bony landmarks of the knee are the patella the two epicondyles of the femur the tibia and the fibula

The patella is pointed distally where the tendo patellae is attached and is slightly convex on its proximal border Its lateral edges are prominent owing to the lack of tissue over this bone With the limb extended and quadriceps muscle relaxed the patella can be moved in all directions except distally There is a depression above the patella When the muscle contracts this depression is filled by the rectus and the muscular swells on each side produced by the vastus internus and externus are visible When the quadriceps muscle is contracted the tense tendo patellae becomes evident when the muscle is relaxed the soft fatty pad beneath the tendon can be felt

About halfway between the patella and the tubercle of the tibia can be felt a groove the line of the joint and the location of the semilunar cartilages On the lateral side posteriorly opposite the level of the tibial tubercle can be felt the head of the fibula Running proximally from the fibula is the tendon of the biceps In front of the biceps can be seen and felt the iliotibial band of the fascia lata It is difficult to distinguish the joint line of the knee by palpation

Posteriorly with the leg extended the condyles of the femur can be outlined and recognized The medial is the more prominent The tubercle of the tibia situated at the insertion of the patellar ligament can best be seen and felt when the tendo patellae is relaxed The tubercle of the tibia is not to be confused with the external tuberosity of the tibia which is proximal and lateral to the tubercle laterally from the external tuberosity and somewhat distally can be seen and felt the head of the fibula Posteriorly when the knee is extended is seen the fullness of the popliteal space

Extending about 2 inches (about 5 cm) above the patella is the subfemoral or suprapatellar bursa

The capsular structures of the knee in their lower ends are attached to the tibial tubercle but above their attachments are far removed from the joint These structures are so strong and thick that pus from within does not come through but goes around them In maximal extension the bulk of the patella rises above the articular surface and connecting its upper edge with the anterior surface of the femur is only the thin capsular ligament hence effusions into the joint bulge upward at this point

Functional Testing of the Muscles About the Knee Joint *Hamstring Muscles* The patient in the prone position tries to flex the knee against resistance The tendons of the biceps (laterally) (Fig 6 57A) and semitendinosus (medially) (Fig 6 57B) * can be felt and usually seen (Nerve supply L_4 L S_1 S Sciatic nerve)

The Knee and Monarticular Disorders ***Infections of the Knee Joint*** The knee joint is affected by both acute and chronic inflammations

In the *acute inflammations* of the knee joint the etiology may be that met with in acute arthritis of any of the joints An acute gonitis is nearly always of infectious nature Gonococcal gonitis is rare it may localize in one knee joint and sometimes

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using the affected extremity as a support The body becomes inclined toward the healthy side

The diagnosis of disease of the hip joint is made from the history and the physical and roentgenologic examinations However, in some instances the exact diagnosis awaits bacteriologic and histologic studies

THE THIGH

The thigh is composed of the femur and three main sets of muscles and is supplied and traversed by the femoral vessels and sciatic and anterior crural nerves The femur serves as a support The muscles move the thigh on the trunk and the leg on the thigh The thigh muscles are extensors flexors and adductors

The deep fascia (fascia lata) of the thigh is a strong layer which completely invests the muscles of the thigh and covers the gluteal region Over the saphenous opening the fascial layer is thin and is known as the fascia cribrosa

The *femoral (Scarpa s) triangle* occupies approximately the proximal third of the thigh Its base proximally is formed by Poupart s ligament its lateral side by the sartorius muscle and its medial side by the adductor longus Its floor is formed by the iliacus psoas pectineus sometimes a portion of the adductor brevis and the adductor longus muscles In the depths of this triangle course the femoral artery and vein the anterior crural nerve the long saphenous vein and numerous lymphatic vessels At its proximal and medial part is the saphenous opening at which femoral hernias make their appearance Psoas abscesses follow the tendon of the psoas muscle down and make their appearance in Scarpa s triangle Also pus from hip joint disease comes to the surface at the upper part of the triangle on one side or the other of the femoral artery The apex of Scarpa s triangle is a favorite site for ligation of the femoral artery

The lymph nodes of the groin are separated into the superficial and the deep sets The nodes of the superficial set are along Poupart s ligament and along the blood vessels The nodes of the groins drain the lower anterior half of the abdomen the genitalia the lower limbs and the anal gluteal and lumbar regions

The deep lymph nodes are not constant and rarely become the seat of infection but when chronically inflamed such a node may be mistaken for a femoral hernia The nodes of the groin communicate with the external and common iliac nodes

Femoral Hernia Femoral hernia although often considered to be acquired is probably the result of a congenital defect The hernia descends through the femoral canal beneath Poupart s ligament to make its appearance at the saphenous opening on the thigh The saphenous opening has its center 1½ inches (about 4 cm) below and to the outer side of the spine of the pubis The femoral canal represents the narrow space between the femoral vein and the inner wall of the femoral sheath Weakness of the ligamentum lacunare (Gimbernat s ligament) which forms the medial boundary of the femoral ring is an important factor in the development of a femoral hernia

Femoral hernia occurs less frequently than inguinal hernia and more often in women than in men

The hernia appears in the soft tissues in the femoral region Such a hernia is smaller than an inguinal hernia and may be present without a mass or other external manifestation or even without the patient being aware of it When strangulation is present and it occurs often outward manifestations may be minimal unless the palpating finger presses on the hernia which is often buried in femoral fat If a loop of bowel is strangulated however the manifestations of intestinal obstruction are obvious

A femoral hernia large enough to contain intestine is obvious Femoral hernias of this size are not common In an obese patient a strangulation of a small femoral hernia may not be diagnosed until operation for intestinal obstruction is performed

A small hard femoral hernia has to be differentiated from an *enlarged lymph node a small lipoma* and *a cirsoid femoral vein* Usually when lymph nodes are enlarged several nodes are affected A small lipoma usually can be grasped in the hand and separated from the deep lying tissue whereas a femoral hernia cannot be

Stiffness of the knee may be extremely persistent following immobilization of fractures of the leg or thigh Permanent stiffening may occur as a result of atrophy of cartilage synovial shrinkage and muscular contraction

Trauma to the knee joint may be followed by loosening of a cartilage from one of the condyles of the femur (roentgenologic examination palpation) or if there has been a twist of the joint especially when the thigh has been rotated outward when the leg is fixed there may be a loosening and displacement of one meniscus usually the medial meniscus This causes a disturbance of function of the joint described as internal derangement

Internal Derangements of the Knee The evidence for internal derangements of the knee is established by the following measures (1) Eliciting the history ascertains the mechanics by which the lesion was made and the amount and type of pain swelling locking or catching instability weakness and limitation of joint motion Many derangements of the knee which seem of little consequence become disabling owing to the rapid atrophy of the quadriceps muscle as a result of injuries of the knee (2) By inspection the degree of atrophy of the quadriceps muscle is seen as well as the range of active and passive motion of the joint the presence or absence of swelling and fluid and the location of tenderness if observed Then tests are made for lateral and anteroposterior instability (3) The roentgenologic examination is usually of value only in a negative way since the majority of internal derangements affect only the soft tissues however it is important in establishing the presence of loose bodies fractures and calcified meniscuses

Internal derangements of the knee may result from (1) injuries of semilunar cartilages such as tears dislocations cysts discoid cartilage and calcified meniscuses (2) loose bodies in the joint such as detached osteophytes synovial osteochondromatosis osteochondritis dissecans and osteochondral fractures (3) injuries of the ligaments (4) affections of the soft tissues such as cysts and adventitious bursae hypertrophic infrapatellar fat pads and intra articular adhesions (5) fractures of the tibial spine and (6) recurrent dislocations of the patella

SEMILUNAR CARTILAGES *Tears of the Semilunar Cartilages* These are usually found in young men The history consists of the triad of pain in the knee swelling of the knee and locking or catching of the knee The triad begins after a twisting injury with the tibia abducted on the femur in case of tears of the medial semilunar cartilage and adducted on the femur in case of tears of the lateral cartilage The medial semilunar cartilage is injured 10 times more frequently than the lateral semilunar cartilage

Hypermobile or Dislocating Semilunar Cartilages These probably constitute a definite entity and on occasion can cause locking but they are rare Hypermobility may be demonstrated by grasping the anterior end of the cartilage and displacing this toward the midline

Cysts of the Semilunar Cartilages These are extremely rare and when present almost invariably involve the lateral semilunar cartilage They can often be palpated on the lateral side of the knee

A Discoid Semilunar Cartilage This represents a congenital defect which usually affects the external semilunar cartilage and is rare This condition is usually manifest clinically early in life and the child complains of a snapping sensation in the knee when walking

Calcified Meniscuses These are sometimes seen in older persons and occasionally cause a popping or snapping sensation This condition can usually be demonstrated roentgenologically

LOOSE BODIES These are the second commonest cause of internal derangement of the knee The common symptom is locking or catching In the majority of cases loose bodies can be demonstrated by anteroposterior lateral or intercondylar notch roentgenograms The bodies may be palpated in the suprapatellar pouch or about the articular surface of the joint Loose bodies may be the result of

it causes suppuration there In polyarthritis due to rheumatic fever the knees are frequently involved

In all acute infections there is evidence of effusion into the knee joint with floating patella usually there is also some thickening of the capsule which can be felt in thin persons at its fold of reflection when the affected knee is compared with the healthy knee Sometimes the cartilages and bones are involved as revealed by roentgenograms

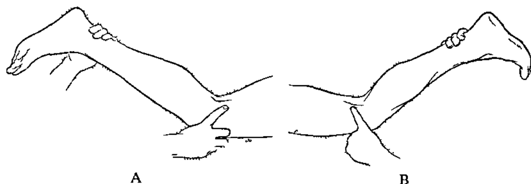


Fig 6 57 Hamstring muscles (semitendinosus and biceps femoris muscles) A laterally biceps muscle B medially semitendinosus muscle Lying on smooth firm surface with face downward patient attempts to flex the knee against resistance The tendon can be felt and seen Nerve supply L_4 L S_1 S (sciatic nerve)

Such acute gonitis must be differentiated from (1) acute prepatellar bursitis (2) hydrops intermittens of the knee and (3) acute osteomyelitis of the lower end of the femur or of the upper end of the tibia

Many different forms of *chronic inflammation* of the knee joint are met with clinically Common causes of chronic effusion into the knee joint are (1) chronic rheumatoid arthritis and (2) tuberculosis

Effects of Trauma In falling the knee often strikes first and there may be an injury to the bursa prepatellaris to the patella or to the knee joint itself

An effusion into the bursa is recognizable from its location and limitations away from the joint When there is an effusion into the knee joint itself the knee cannot be fully flexed and there is swelling around the patella the grooves on both sides of this bone being filled out There may be a transverse swelling above the patella where the joint cavity extends up beneath the tendon of the quadriceps muscle Effusion into the joint lifts the patella from the condyles of the femur If pressed with the finger down upon the bone the firm substratum can be felt when it is reached if the pressure is taken off again the patella will once more float

In a sprain some of the ligamentous or cartilaginous structures may become permanently damaged The lateral ligaments are the more commonly injured The internal lateral ligament is more commonly injured than the external and the manifestations may resemble that of injury to the semilunar cartilage The sprain may be severe enough to produce a tear into the bony tissue and in such instances longer and more thorough immobilization will have to be instituted and thus stiff ness of the joint is more likely to occur

Other results of injury to the knee are those of rupture of the crucial ligaments or tearing of the synovial membrane so as to leave a fringe that may be caught in joint action and create chronic synovitis

True dislocation of the knee joint is rare and is produced by extreme violence It is common however for the patient to believe that the knee has been dislocated following some slipping or catching sensation due to a strain or a sprain True dis location may be forward backward or lateral and may be associated with fracture

tendon Behind the knee joint in the popliteal space or poples swelling due to aneurysm is not uncommon its pulsation quickly differentiates it from (1) distended bursa here (2) lipoma and (3) cold abscess

Of the tumors below the knee sarcoma of the upper end of the tibia or of the fibula is not uncommon The roentgenogram in such cases is characteristic Cartilaginous exostoses may also occur near the extremities of both the tibia and the fibula

THE LEG

The two tuberosities of the tibia the tibial tubercle and the head of the fibula can be seen or palpated

The tibia is triangular in shape with a sharp edge—the crest or shin—forward and a flat surface medially Thus it participates in the formation of two surfaces the medial and the anterior of the leg The medial surface is subcutaneous and extends almost the full length of the leg to the internal malleolus The lateral surface has the extensor muscles between it and the fibula The fibula a short distance below its head becomes covered by the peronei muscles and becomes subcutaneous only in its lower anterior fourth The proximal portion of the leg is largely muscular but at its distal portion it is mainly tendinous

The muscles of the leg can hardly be considered separately from those of the foot The long muscles support the arch of the foot flex extend abduct and adduct the foot Adduction of the foot is associated with a limited amount of rotation

The individual muscles perform compound action not a simple one They act on two joints the ankle and subastragaloid If the ankle joint is stationary they abduct and adduct the foot If the subastragaloid joint is stationary they flex and extend the foot but if both joints move as is usually the case combined action of the muscles accomplishes this feat

The Action of the Muscles as Flexors and Extensors The movements of the foot when strength is not required are performed by the flexor and extensor groups of muscles The powerful calf muscles aid especially in lifting and propelling the body forward in locomotion When most of the flexors and extensors of the leg are paralyzed the foot hangs loose from the leg the so called flail foot (drop foot) Weakness of the flexor group (tibialis posterior flexor digitorum longus and flexor hallucis longus) tends to favor a descent of the arch with consequent pronation or eversion Weakness of the extensors causes toe drop and inversion or supination

Paralysis of the calf muscles deprives the posterior pillar of the arch of its support and the action of the flexors and extensors elevates the arch while the heel descends so that a condition of hollow foot is produced

The Action of the Muscles as Abductors and Adductors Lateral movements of the foot are comparatively weak These movements are for maintenance of equilibrium and adaptation of the position of the foot to uneven surfaces Three muscles act very distinctly as abductors they are the peroneus longus brevis and tertius Two act as distinct adductors namely the tibialis anterior and the tibialis posterior

The muscles of the calf act more as abductors than adductors because the insertion of the Achilles tendon (tendo calcaneus tendo achillis) is not directly behind the ankle joint but more to its outer side

When the foot is deformed in the position of inversion as in clubfoot the tibialis anterior and posterior are usually contracted but when in the position of eversion as in flatfoot spasm of the peronei or calf muscles is frequent

Plantar flexion of the foot is a far more powerful movement than extension Flexion is associated with adduction or inversion and extension with abduction or eversion hence it is that inversion is the position of strength and eversion of weakness Feats of strength and agility cannot be performed by those who have markedly everted feet

Detached Osteophytes These occur in persons of later middle age or senility who have had osteo arthritis for years

Synovial Osteochondromatosis There are numerous loose bodies from the synovial membrane

Osteochondritis Dissecans This condition usually begins between 10 and 19 years of age and probably represents a form of aseptic necrosis of subchondral bone The loose piece of subchondral bone with its overlying hyaline cartilage may not be present in the joint as a true loose body and yet the patient may have considerable distress in the form of a dull aching pain The roentgenogram of the intercondylar notch is often invaluable in demonstrating this lesion

Osteochondral Fracture This type of loose body results from a fracture involving the joint surface whereby a loose fragment is displaced into the joint

AFFECTIONS OF THE SOFT TISSUES *Cysts and Adventitious Bursae* These some times appear about the ligaments of the knees and may cause pain and disability of the knee

Hypertrophic Infrapatellar Fat Pads These sometimes occur but are probably very rare This condition may cause a locking or catching sensation as the fat pad is pinched between the articular surfaces of the femur and the tibia

Intra Articular Adhesions These sometimes limit the range of motion of the joint or may even cause a catching sensation but rarely a true locking

FRACTURES OF THE TIBIAL SPINE These really are lesions of the cruciate liga ments A positive diagnosis can be made only by roentgenologic methods and espe cially by a roentgenogram of the intercondylar notch

RECURRENT DISLOCATION OF THE PATELLA This lesion is much more often encountered in women than in men and is often on a congenital basis The patient gives a history of acute episodes characterized by a sudden slipping and locking sensation with severe pain The knee may remain locked in flexion If the patient is observant she will state that the kneecap seems to be displaced to the lateral side of the knee and that she usually manipulates it back to its original position where upon the acute pain rapidly disappears The knee after this episode becomes con siderably swollen and may be moderately painful for 1 to 2 weeks

Ankylosis of the Knee Joint The knee joint is of the hinge type but has a slight freedom of action in rotation and lateral mobility so that it is also a gliding joint

In ankylosis of the knee rigid fixation in extension is more favorable to function while flexion is less favorable However slight flexion has an advantage since by shorten ing the foot has more room to pass forward along the ground

Contusion to the knee joint may produce synovitis followed by adhesions and limita tion of motion

Joint swelling of the knee or synovitis with local redness and heat associated with fever is an infectious process not an injury Trauma does not produce the symptoms of inflammation as does infection Synovitis of the knee produced by a strain or sprain causes swelling and distention of the joint but does not cause fever or a generally reddened hot painful joint

In either partial or complete ankylosis of the knee the nearer the motion can approach full extension the less the disability Fifteen or 20 degrees of flexion starting from full extension permits most of the activities of the leg as a support and walking can be carried out with little mechanical interference (McBride)

Tumors and Swellings Of the tumors in the region of the knee joint lipoma is the commonest Occasionally a sarcoma of one of the bones is met with or a fibroma or sarcoma may originate in the synovial membrane

Chronic swellings in the neighborhood of the knee joint other than neoplasm are commoner Thus in front of the joint swellings are due to (1) chronic bursitis prepatellaris (2) chronic bursitis pretibialis or (3) bursitis behind the patellar

Peroneus Longus and Brevis The patient tries to evert the foot against resistance The tendons can be felt and usually seen (Fig 6 65) (Sciatic nerve supply L S_1)

Fig 6 61 Tibialis anterior muscle Lying on back or sitting patient attempts to dorsi flex the toes against resistance The tendons can be seen and felt Nerve supply L_4 L S_1 (sciatic nerve)

Fig 6 62 Extensor digitorum longus muscle Lying on back or sitting patient attempts to dorsiflex the toes against re sistance The muscles can be seen and felt Nerve supply L_5 S_1 (sciatic nerve)



Fig 6 61



Fig 6 6-

Small Muscles of Foot The patient tries to make a cup of the sole of the foot (Fig 6 66) (Sciatic nerve supply S_1 S) *

Anterior Tibial Syndrome The anterior tibial syndrome occurs chiefly in young men and consists of ischemic necrosis of the muscles of the anterior tibial

Fig 6 63 Extensor hallucis longus muscle Lying on back or sitting patient attempts to dorsiflex the great toe against resistance The tendon can be felt and seen Nerve supply L S_1 (sciatic nerve)

Fig 6-64 Extensor digitorum brevis muscle Lying on the back or sitting patient attempts to dorsiflex the great toe against resistance The muscle can be seen and felt Nerve supply S_1 (sciatic nerve)



Fig 6 63



Fig 6 64

* Acknowledgment is made to the Controller of Her Britannic Majesty s Stationery Office for permission to make line drawings of photographs of muscles in action in War Memorandum No 7 reprinted 1945

Functional Testing of the Muscles of the Leg and Foot *Gastrocnemius* The patient in the prone position tries to plantar flex the ankle joint against resistance The muscle belly and tendon can be seen and felt (Fig 6-58) (Sciatic nerve supply, S_1 S)

Tibialis Posterior The patient lies on the back and tries to invert the plantar flexed foot against resistance The tendon can be seen and felt (Fig 6 59) (Sciatic nerve supply L_4 L)



Fig 6 58 Gastrocnemius muscle Lying on belly or on side with legs extended patient attempts against resistance to plantar flex the ankle The muscle can be seen and felt Nerve supply S_1 S (sciatic nerve)



Fig 6 59 Tibialis posterior muscle Lying on back with legs extended and abducted the foot plantar flexed patient attempts to invert the foot against resistance The tendon can be seen and felt Nerve supply L_4 L_5 (sciatic nerve)



Fig 6 60 Flexor digitorum flexor hallucis longus muscles Lying on back legs abducted and extended and the foot plantar flexed patient attempts to flex the terminal joints of the toe against slight resistance The muscles can be felt Nerve supply S_1 S (sciatic nerve)

Flexor Digitorum Longus Flexor Hallucis Longus The patient tries to flex the terminal joints of the toes against slight resistance The muscle bellies can be felt (Fig 6 60) (Sciatic nerve supply S_1 S)

Tibialis Anterior The patient tries to dorsiflex the ankle against resistance The muscle belly and its tendon can be seen and felt (Fig 6 61) (Sciatic nerve supply L_4 L S_1)

Extensor Digitorum Longus The patient tries to dorsiflex the toes against resistance The tendons passing to the outer four toes can be seen and felt (Fig 6 62) (Sciatic nerve supply L_5 S_1)

Extensor Hallucis Longus The patient tries to dorsiflex the great toe against resistance The tendon can be seen and felt (Fig 6 63) (Sciatic nerve supply L S_1)

Extensor Digitorum Brevis The patient tries to dorsiflex the great toe against resistance The muscle belly can be felt and often seen (Fig 6 64) (Sciatic nerve supply S_1)

condition is worse when the patient is resting then there is an uncontrollable desire to move the legs Generally speaking continuous paresthesia points to an organic lesion of the nervous system while intermittent paresthesia of the type described in this syndrome points to a functional disorder possibly of vasomotor nature

Massage has no effect but getting up and walking about may give relief The disagreeable sensation stops at once on chewing 1/100 grain of nitroglycerin

Fat Legs and Edema (Lipedema) Those who have lipedema complain of en largement of the limbs aching distress in them particularly during activity and marked tenderness of the legs The emotional reaction of patients to this syndrome varies from curiosity relative to its significance to marked evidence of anxiety and tension In women who are desirous of maintaining a youthful appearance the emo tional reactions to the fat legs may be considerable There are insomnia nervous ness tenseness melancholia anxiety and feelings of frustration Such a woman is ashamed of her legs or feels that her large legs have ruined her life These feelings and symptoms are understandable reactions to a disfigurement which is in conflict with the shapely legs of the model for hosiery advertisements Understandable too is the wearing of long skirts to hide the legs the avoidance of appearing in swimming suits and the standing behind chairs at parties

Characteristically there is symmetric bilateral enlargement of the buttocks and lower extremities which begins almost imperceptibly and progresses gradually Progressive enlargement of the limbs is ordinarily associated with gain of weight but evidence of obesity of the trunk upper extremities face and neck may be entirely absent in some instances there is generalized obesity The enlargement of the limbs is accentuated by orthostatic activity particularly in warm weather Although rest in bed may cause some decrease in size of the legs it will not cause the limbs to return to a normal size Recurring inflammations such as are observed in lymphedema are uniformly absent The feet are normal in size and configuration There is moderate to great sensitiveness to digital pressure

THE ANKLE

The ankle is a hinge joint and its motion is anteroposterior except in maximal exten sion when a limited degree of lateral movement is possible The range of movement of the ankle is 80 degrees 20 degrees of flexion and 60 degrees of extension

The contour of the ankle is rather uniform but this uniformity is easily destroyed by diseases and injuries The tibial malleoli form prominences with distinct hollows above and below them The sharp anterior edge of the tibia if followed distally leads to the tibialis anterior tendon On the medial (inner) side the malleolus is large and flat It is subcutaneous and can be readily palpated At its anterior edge is the com mencement of the long saphenous vein which runs proximally and slightly back to reach the posterior edge of the tibia About 1½ inch (about 4 cm) distal and in front of the internal malleolus is the prominent tubercle of the scaphoid

The external malleolus is small and somewhat pointed and is placed a finger breadth distal and behind the level of the internal malleolus Proximal to the tip of the external malleolus is the fibula which is subcutaneous and readily palpated The transverse line of the joint is level with the upper limit of the swell of the internal malleolus The ankle is covered in front and behind by tendons most of which especially in thin persons can be felt and seen when their muscles are contracted

Posteriorly the tendo calcaneous (tendo achillis) is large and prominent The posterior tibial artery can be felt pulsating midway between the tendo calcaneus and the internal malleolus The anterior tibial artery can be felt pulsating to the lateral (outer) side of the extensor hallucis longus

Sprain of the Ankle In what is usually called a sprain of the ankle the injury is not always confined to the ankle joint and its ligaments In many cases there is a

portion of the leg with a lesion of the anterior tibial nerve The onset of symptoms is directly related to trauma or physical exertion involving strenuous use of the leg muscles Unaccustomed exertion causes muscle trauma which is followed by increased pressure within the anterior tibial region impaired blood supply to the affected muscles and ischemic necrosis *The anterior tibial nerve* is involved either by compression, in which case it recovers rapidly or by ischemia in which case the loss of function is permanent The symptoms are pain in the front of the leg fol

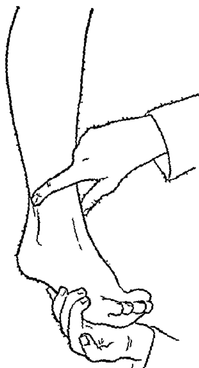


Fig 6 65 Peroneus longus and brevis muscles Lying on back with legs extended and abducted patient attempts to evert the foot against resistance Tendons can be felt and seen Nerve supply L S_1 (sciatic nerve)



Fig 6 66 Small muscles of sole of foot Lying on back with legs extended patient attempts to cup the sole of the foot Nerve supply S_1 S (sciatic nerve)

lowed by signs of inflammation over the pretibial muscles and inability to dorsiflex the foot and toes Important negative observations are normal action of the peroneal muscles and a minimal degree of foot drop If the syndrome is recognized early rest alone may prevent irreversible damage to the muscles

Restless Legs (Anxietas Tibiarum Leg Jitters) The symptoms are subjective and consist of paresthesias a feeling of weakness in the legs and a sensation of cold in the feet The paresthesias occur over the inner aspect of the legs sometimes in the thighs but rarely in the feet Characteristically the crawling sensation develops only when the legs are *still most often after the patient retires for the night Sleep* may be disturbed night after night for years with periods of improvement and exacerbation The disease or syndrome occurs in two main forms one is characterized mainly by paresthesia and is termed asthenia crurum paresthetica by Ekbom and the other in which pain predominates is termed asthenia crurum dolorosa

The paresthetic variety is an easily recognizable condition probably closely related to acroparesthesia and nocturnal burning feet The painful variety consists of mild to moderate aching localized in the same situations as the paresthesia The

necessitate more work for the long and short muscles and tend to cause foot tire and leg ache

When all of the weight is borne on the ball of the foot as in walking on tiptoe it is called toe to toe walking Toe to toe walking imparts greater elasticity and spring to the gait and results in less shock impact than heel to-toe walking Because of the added elasticity and spring which is secured by toe to toe walking it is used in running and because it is more graceful it is used in dancing

The two tubercles of the calcaneous (os calcis) can be felt posteriorly and at the sides The external surface can be followed forward but the internal is buried be neath the soft tissues Of the two tubercles on its undersurface the internal can be felt on the sole of the foot by firm pressure

The tubercle of the scaphoid (navicular) lies on the plantar rather than on the lateral aspect of the bone It can be felt 1½ inches (about 4 cm) below and in front of the internal malleolus It is the landmark for the tarsal joints on the inner side of the foot The tibialis posterior muscle runs from it to the posterior edge of the internal malleolus

The tuberosity of the fifth metatarsal bone is the large bony prominence 2½ inches (about 6 cm) below and in front of the external malleolus It is the guide to the tarsal joints on the outside of the foot The tendon of the peroneus brevis runs from it to the posterior edge of the external malleolus

ANKYLOSIS OF THE TOE JOINTS

Complete Ankylosis Fixed ankylosis of the big toe at the metatarsophalangeal joint is termed hallux rigidus this is the commonest site of complete ankylosis This condition may arise as the result of osteoarthritic changes or from severe fracture ex tending into the joint It may also result from infection and occasionally from operations which involve the joint When ankylosis occurs a very slight dorsiflexion is of advantage

The middle phalangeal joint may also become rigidly ankylosed but does not have the importance of the metatarsophalangeal joint The lesser toes may be individually or collectively ankylosed If the metatarsophalangeal joints are fixed in a dorsiflexed posi tion there occurs a prominence to the ball of the foot so that the anterior arch is obliterated The condition is termed clawfoot

When the middle phalangeal joint becomes ankylosed in a flexed position the meta tarsophalangeal joint is usually dorsiflexed and the condition is termed hammer toe

Partial Ankylosis Partial limitation of the ankle and tarsal joints is one of the commonest disabilities of the foot Particularly the plantar flexion deformity is common because so frequently injuries of the leg or foot are treated with the foot in a splint which does not hold it to a right angle

The common causes of partial ankylosis of the toes are (1) contusions or sprains (2) dislocations (3) fractures in or near the joints (4) continuous immobilization (5) scar contracture (6) suppuration and (7) rheumatic arthritis

When a toe is examined a considerable length of time after the healing of an injury the amount of movement that was normal for the toe previous to injury is unknown except by comparison with the movements of the toes of the other foot On account of the confinement of the toes within the shoe there is a variation in the shape and motion of toes

ANATOMIC DERANGEMENTS AND FUNCTIONAL DISORDERS OF THE FOOT

Foot Imbalance Children have one basic type of foot imbalance—*pes planus* The adolescent has two basic types—*pes planus* and *pes cavus* The adult has both of these imbalances and as the result trouble may originate in the anterior arch

Pes Planus (*Flatfoot*) A flat foot is characterized by inrolling or pronation of the foot and a depression or lowering of the longitudinal arch

tearing off of small fragments of bone hence the name sprain fracture Inasmuch as sprains are usually the consequences of lateral displacement the resultant injury is frequently in the subastragaloid and sometimes in the adjacent tarsal joints A sprain is frequently the result of an inversion rather than of an eversion of the foot because in eversion the plantar ligaments are so strong that the foot moves as a whole and the force is transmitted directly to the ankle and leg bones

A sprain of the ankle is accompanied by pain swelling and loss of function but there is no subcutaneous ecchymosis

Ankylosis of the Ankle Joint The ankle joint is a hinge joint with flexion and extension A wide range of motion is provided by the tarsal joints The muscle relationship between the ankle and tarsal joints is correlative

The common causes of partial ankylosis are contusions strains sprains fractures (Pott's fracture) dislocations continuous immobilization scar contractures, suppurations and rheumatic arthritis

When the foot cannot be brought to the right angle position, the toe drags as the foot advances Toe drag creates awkwardness and stumbling Strength security and co ordination are most affected but quickness and endurance are also decreased to a degree proportional to the extent of equinus An individual who must walk on tiptoe has a noticeable deformity and limp

When plantar flexion of the foot is limited to a right angle or less the foot cannot participate in the acts of stepping running or jumping Equilibrium becomes unstable The foot is strong however and co-ordination and endurance are not greatly affected so long as the foot can reach a right angle position

When the degree of abduction or pronation is limited to the midline there is interference in shifting of the weight in taking a step The gait in the presence of eversion is limited in smoothness and there may be loss of endurance In squatting or jumping there is need of abduction to permit complete dorsiflexion of the foot Limitation of eversion however does not prohibit use of the foot for any particular activity When abduction and eversion are limited to the extent that the foot cannot be brought to the midline the deformity and limp are obvious

When the foot is everted and cannot be adducted there is an increased strain on the arch and endurance is affected The foot lacks the flexible inward rotation and the weight must therefore be borne mostly on the big toe in taking a step or while standing (McBride)

THE FOOT

The foot provides support for the weight of the body acts as a lever to raise and to propel it into motion and when the body is in motion acts to prevent it from receiving sudden or unexpected jolts

If standing is too prolonged ligaments stretch and permit abnormal separation of the foot bones and induce a true foot strain The standing position in which the foot points straight forward or in which there is moderate outtoeing is the position in which locking of the bones of the foot is at its maximum and the ligaments are under the least strain

With the body in motion the muscles play a more important role in supporting the foot than they do with the body at rest because of the element of postural stability The long muscles become more important as supports of the arches since by maintaining the leg in balance over the foot they prevent abnormal concentration of weight stresses on one part of the foot

In heel to toe walking to be efficient the foot points directly forward in the sagittal plane with each step or at least toes out only 20 to 30 degrees The foot in the adducted position is from a structural point of view in a stronger position than that in which the foot is in the neutral or straight position When the foot toes out more than 30 degrees the loss of flexibility and interference with smooth action

normal range of dorsiflexion of the first metatarsal bone indicates a hypermobile first metatarsal segment Callosities under the heads of metatarsals I II and V and at times over the entire ball of the foot are frequently observed

In the rigid flat foot the tendons of the peroneus longus and brevis are short and contracted The lateral mobility of the foot is limited

The roentgenogram may reveal a short first metatarsal with overdevelopment of the second metatarsal a metatarsus varus primus with wide separation between metatarsals I and II a hypermobile first metatarsal segment or arthritic changes in the foot joints

DIAGNOSIS The diagnosis of pes planus is based on the foregoing symptoms and objective findings

Pes Cavus (*High Arched Foot*) Pes cavus is often congenital but may occur as the result of muscle imbalance secondary to muscle paralysis especially owing to infantile paralysis when it is almost invariably due to weakness of the dorsal flexors of the toes and overaction of plantar flexors plantar intrinsic muscles and heel tendon Pes cavus also occurs in spastic paralysis due to overaction of muscles

SYMPTOMS There is pain in the ball of the foot described as a feeling of walking on something hard under the ball of the foot There is tiring in the longitudinal arch of the foot particularly across the dorsum and cramping in the calf of the leg Backache is often present if anteroposterior balance is disturbed by a tilting of the pelvis downward and forward

There is less flexibility in the foot as a whole than in a normal or a flat foot

EXAMINATION With the patient in the standing position inspection will reveal that the longitudinal arch is abnormally high the forepart of the foot is somewhat adducted and the toes are in There may be the appearance of pronation owing to inward rolling of the foot at the ankle joint The line of transmitted weight falls toward the lateral side of the foot which tends to roll it outward

When the patient sits it is observed that the Achilles tendon is short either actually or relatively because of dropping of the forepart of the foot The ball of the foot is prominent and there is callus formation Tenderness is present over the contracted plantar fascia and over the heads of the metatarsal bones

DIAGNOSIS The diagnosis of pes cavus is based on the presence of an abnormally high arch shortening of the plantar fascia prominence of the ball of the foot and callus formation There is an absence of pronation and there is shortening or at least functional shortening of the Achilles tendon Roentgenograms yield very little information

Descent of the Metatarsal Arch and Anterior Metatarsalgia A depression of the metatarsal arch is associated with either pes planus or pes cavus Disturbances of the metatarsal arch may occur as a separate entity in women more than 30 years of age who have worn improperly fitting shoes

A depressed metatarsal arch is due to improper distribution of weight stresses over the foot as the result of pes planus pes cavus and improper footwear High heeled pointed narrow toed shoes displace the burden of weight bearing on the metatarsal arch and limit the forepart of the foot and toes in their normal movements

There is discomfort in the ball of the foot often amounting to acute cramplike burning pain in the second and fourth toes There is loss of the normal dorsal convexity of the metatarsal arch The toes are flexed so that a hammer toe position is assumed Callus formation often extends across the ball of the foot

Metatarsalgia (*Morton s Toe*) Anterior metatarsalgia represents an advanced form of a depressed metatarsal arch Anterior metatarsalgia is characterized by the sudden onset while walking of an acute burning pain in the fourth toe but at times in the second toe This pain is so severe that the shoe must be removed and the toes

ETIOLOGY Flat feet may result from (1) deformities of the bone (2) short Achilles tendon (tendo calcaneus heel cord) (3) muscle inadequacy (4) ill fitting shoes and (5) knock knees bowlegs and tibial torsion

Deformities of the bones constitute a short metatarsal I metatarsus varus primus hypermobility of the first metatarsal segment and accessory scaphoid or prehallux in addition to those resulting from trauma and infection which are so numerous as to be indescribable The architecture of the foot may be deformed from fracture of bones and loss of the great toe

Hallux valgus develops by depriving the medial side of the foot of the supporting action of the great toe allowing the foot to pronate which encourages the descent of the longitudinal arch Fractures of the os calcis commonly cause a depression of the longitudinal arch

A short Achilles tendon prevents dorsiflexion of the foot and shifts the weight bearing to the anterior part of the foot The weight on the forepart of the foot causes downward and inward displacement of the subastragalar joint and descent of the longitudinal arch The pull of a short Achilles tendon tends to displace the os calcis inward and to tilt the subastragalar joint downward and inward which depresses the longitudinal arch of the foot

Flatfoot may be due to an inability of the muscles to maintain the leg in an approximately vertical plane over the foot Weakness of the leg and foot muscles may result from illness obesity or muscle paralysis A commoner cause of muscle weakness is prolonged standing usually occupational

Shoes having high heels and inadequate length and width may cause foot imbalance In such shoes the toes are crowded into the forepart of the shoes and thus the weight is borne on tiptoes

Normally the line of transmitted weight of the body passes approximately through the middle of the patella straight downward between metatarsals I and II In the presence of knock knee the line of weight bearing is shifted through metatarsal I or medial to it With bowleg and tibial torsion the line of weight bearing falls through metatarsal I or medial to it just as it does in knock knee and the weight bearing thrust is on the medial side of the foot Excessive weight bearing here rotates the medial side of the foot downward and inward at the subastragalar joint with eventual depression of the longitudinal arch

SYMPTOMS The first symptom is a tired feeling in the longitudinal arch and the calf of the leg This tired feeling gradually increases in intensity and in time becomes a disabling pain The pain is localized in the region of the scaphoid bone and the subastragalar joint but may include the entire foot Cramping pain in the calf of the leg is often present Pain develops in the knee as the result of the shifting inward of the line of transmitted weight

A downward tilting of the front of the pelvis accompanied by the secondary upward and forward tilting of the back of the pelvis which may result from extreme instances of inrolling of the feet increases the work of the lumbar muscles and ligaments and is said to be responsible for the backache in pes planus

EXAMINATION When making an examination for objective evidence of pes planus or flatfoot request the patient to stand with the feet parallel and about 3 inches (about 8 cm) apart If there is flatfoot the longitudinal arch is depressed The foot is pronated so that the line of transmitted weight falls through metatarsal I or even medial to the inner border of the foot

The scaphoid bone and the internal malleolus are prominent owing to pronation or downward and inward rolling of the subastragalar joint The great toe is short compared with the other toes Prominence of the great toe with lateral projection and a broad splayed out forefoot are present in metatarsus varus primus

Tenderness to palpation is usually present in the region of the scaphoid bone and the head of the astragalus and over the plantar surface of the middle cuneiform bone

The range of dorsal flexion in the male foot with the knee extended and the foot held in mild adduction is 85 or 90 degrees in the female foot 90 degrees An ab

and the tip of the toe impinges on the ground it is called mallet toe This toe deformity gives remarkably little distress in most instances

Congenital hammer toe often involves the second or fifth toe it is often a familial characteristic but may be observed when the foot is paralyzed

In rare instances the deformed toe is affected by bursitis In mallet toe a sensitive callosity may form over the end of the toe in close proximity to the nail

Injuries to the Sesamoid Bones The sesamoid bones of the foot are often irritated injured and occasionally fractured Improper weight bearing may cause a localized irritation of the sesamoid bones of the great toe joint Proliferative changes may take place in the sesamoids which increase in size and become irregular in shape exostoses may even be felt about the margins of such an irritated sesamoid Irritation of a sesamoid is characterized by local pain over the involved bone and tenderness at the same point

Affections of the Tarsal and Metatarsal Bones ***Köhler s Disease of the Tarsal Scaphoid*** Kohler s disease occurs in children between the ages of 4 and 7 years and is found more frequently in boys than in girls The cause is a degenerative bone condition which occurs in the young Trauma is a contributing causative factor Necrotic changes in the cancellous bone with replacement fibrosis have been found

There is localized pain over the scaphoid bone aggravated by use and accompanied by a limp Objectively there is tenderness over the scaphoid bone and at times swelling Roentgenograms taken in the dorsoplantar and lateral planes show that the tarsal scaphoid is small dense often irregular and narrow in its anteroposterior diameter

March Foot March foot is characterized by a painful swelling on the dorsum of the foot often associated with a spontaneous fracture of one of the metatarsal bones usually the second or third The condition follows long walks with excessive weight on the back

Interference with the blood supply with weakening of the bones has been suggested as a cause of march foot Extensive use of the forepart of the foot under adverse conditions is the most logical explanation of the condition

When spontaneous fracture occurs there are periosteal proliferation fracture of the involved metatarsal bone and finally as healing progresses excess callus formation

There is pain in the fore part of the foot Walking is seriously interfered with or is impossible There is swelling over the dorsum of the foot with localized redness and pain There is an area tender to pressure over the distal part of the second or third metatarsal bone If fracture is present it is impossible to bear weight on the foot After the lapse of a week or two a roentgenogram will show a periosteal fuzziness at the site of the beginning fracture

Infraction of the Second Metatarsal Bone (*Freiberg s Disease*) This is an affection of the head of the second metatarsal bone occurring in adolescents and characterized by degenerative changes with pain in the metatarsophalangeal joint of the second toe Trauma plays a contributing role in the causation of the condition

The head of the second metatarsal bone becomes distorted and irregular from degenerative changes There is an accompanying thickening of the shaft of the bone Pain is present over the head of the second metatarsal bone and walking is interfered with There are localized swelling and thickening of the metatarsophalangeal joint of the second toe The joint is tender to pressure and may show evidence of inflammatory reaction Roentgenograms taken in the dorsoplantar plane show characteristic changes in the head of the second metatarsal bone

Affections of the Heel ***Calcaneal Spurs*** Proliferative changes over the tuberosity of the os calcis at the attachment of the plantar fascia may develop into spurs or spurlike prominences Formerly calcaneal spurs were erroneously considered to be caused by a neisserian infection However systemic infection local irri

massaged and manipulated to relieve the discomfort Attacks of pain tend to become more and more frequent and the duration more prolonged

The digital nerves come to lie in such a position that they are pressed on There may or may not be a tumor present which increases the pressure on the nerve The tumor often present in Morton s toe is either a neurofibroma or an angioneuro fibroma The nerve pressed on in metatarsalgia is as a rule the fourth plantar nerve which is firmly held down against excursions of the toe by the short flexor of the fourth toe producing conditions favorable to trauma

Congenital Clubfoot Congenital clubfoot occurs about once in each 1 000 births A hereditary background is ascertained in 5 to 45 per cent of the cases There are four deformities present in congenital clubfoot (1) adduction of the forepart of the foot (2) inversion of the hind part of the foot (3) equinus and (4) tibial torsion Hart has stated that most children with a congenital clubfoot deformity treated within the first 6 months of life can obtain a functionally normal foot by nonoperative treatment

Hallux Hallux Valgus (*Bunion*) Hallux valgus is characterized by lateral angulation of the great toe at the metatarsophalangeal joint and enlargement of the medial side of the head of the first metatarsal bone with the formation of a bony prominence which in time becomes covered by a bursal sac

The basic cause of hallux valgus lies in a weakness of the architecture of the foot There is often a hypermobile first metatarsal segment and there are depressions of the longitudinal and metatarsal arches

Degrees of hallux valgus are commonly present without symptoms Burning pain at the metatarsophalangeal joint of the great toe is common among those who have symptoms The pain is caused by pressure and irritation of the prominence

There is a lateral deviation of the great toe and the prominence on the medial side of the head of the first metatarsal as stated and often a hammer toe deformity of the second toe is present Depression of the longitudinal arch due to the loss of support of the great toe is often present The metatarsal arch is often depressed and there is callus formation over the head of the second metatarsal bone The bursa over the prominence often becomes distended with fluid and inflamed and even infected when it may break down with sinus formation

Hallux Varus Hallux varus is a congenital deformity in which the great toe is smaller than normal and is held projected toward the medial side of the foot it is usually associated with metatarsus varus

Tailor s Bunion A bunion over the tuberosity of the fifth metatarsal bone has been called a tailor s bunion because tailors often have this type of bunion owing to the position in which they sit while sewing The condition is largely due to friction over the prominent tuberosity of the fifth metatarsal bone by the outer side of the shoe

Hallux Rigidus (*Hallux Flexus*) Hallux rigidus involves the metatarsophalangeal joint of the great toe There is limitation of motion in this joint It may be associated with hallux valgus Ununited phalangeal fractures cause hallux flexus

Stumping trauma to the great toe joint and faulty weight bearing cause this condition It is often a manifestation of arthritis

Stiffness preventing flexion in the great toe causes foot strain and pain results on walking The movements of dorsal and plantar flexion are limited The toe may become fixed in plantar flexion A ridge of bone is palpable on the dorsum of the great toe joint Roentgenograms taken in the dorsoplantar and lateral planes will show narrowing of the joint space and hypertrophic changes in the head of the first metatarsal bone and the base of the first phalanx

Hammer Toe Hammer toe often developing early in life is a deformity of one or more toes characterized by acute plantar flexion and rigidity of the midphalangeal joint and extension at the distal phalangeal joint If the distal joint remains straight

Club Nail (Onychogryposis) Club nail or onychogryposis is an excessive hypertrophy of the nail The nail of the great toe is most frequently affected

The twisted and thickened nail is generally found in the aged and is thought to be due to an irritation or pressure on the nail end which in turn irritates the matrix causing the nail to grow unevenly and at an increased rate The dorsal layers grow more profusely than do the plantar layers hence the end of the nail curls downward and the dorsal lavers pile up to form a hornlike structure

The treatment consists in clipping the nail short with heavy bone shears This will give temporary relief but the overgrowth of nail generally continues It is often advisable to soften the nail with salicylic acid or calcium sulfide before clipping When the club nail is sufficiently enlarged it should be removed

DISEASES COMMON TO THE AXIAL AND APPENDICULAR REGIONS AND AFFECTING LOCOMOTION

DISEASES AFFECTING MULTIPLE JOINTS AND LOCOMOTION

The disability in joint disease depends to some extent on the nature of the disease and the kind or class of joint affected Three classes of joints are described namely fibrous cartilaginous and synovial

Movements About Joints Bones connected by the various types of joints are capable of the following different kinds of movement

Gliding movement is the simplest kind of motion that can take place in a joint one surface gliding or moving over another without any angular or rotatory movement

Angular movement occurs only between the long bones and by it the angle between the two bones is either increased or diminished It includes flexion extension abduction and adduction

Flexion is ventral bending except at the knee joint while in the four footed position

Extension is straightening out of bones of a joint hence the reverse of flexion

Abduction means to draw away from the middle line of the body

Adduction means brought to or nearer the middle line of the body

Both abduction and adduction have different meanings when used with reference to the fingers and toes In the hand the imaginary line is supposed to be drawn through the middle finger and in the foot through the second toe

Circumduction is that form of motion which takes place between the head of a bone and its articular cavity when the bone is made to circumscribe a conical space by rotation around an imaginary axis for instance swinging the arms or legs

Rotation is a form of movement in which a bone moves around a central axis often an imaginary one without undergoing any displacement from this axis No part of the body is capable of perfect rotation like a wheel for the simple reason that such motion would necessarily tear asunder all the vessels nerves and muscles which unite it with other parts

Joint Pain Pain is experienced when any of the sensitive structures of the joint are stimulated with a faradic or direct current or by pinching tearing burning with a cautery or acid cutting with a knife or chisel or sticking with a needle Thus the *structures of a joint are all sensitive* to acute injury

A painful stimulation or injury of fascia tendons and periosteum according to McEwin gives rise to pain felt at the corresponding site of injury distal to it

The stimuli which originate pain in traumatic lesions and in such derangements of a joint as occur in osteoarthritis are likely due to mechanical tearing and squeezing which stimulate the nerve endings and secondarily give rise to inflammatory edema which presses on nerve endings and exerts tension on the nerve networks In infections of the joints inflammatory edema and toxic or chemical irritants released as part of the inflammatory process are sufficient in themselves to cause joint pain Less is known about the mechanism of pain production in *periarticular painful conditions* such as the arthralgias and fibrositis

tation caused by faulty foot attitude and acute or repeated trauma are etiologically important

There is pain in the heel over the tuberosity of the os calcis aggravated by weight bearing There is a definite point of acute tenderness over the tuberosity of the os calcis A lateral roentgenogram will show proliferative changes over the tuberosity of the os calcis and frequently but not always a true spur formation

Calcaneal Apophysitis (*Epiphysitis of the Os Calcis*) The epiphysis of the os calcis lies at the tip of the heel and receives the attachment of the Achilles tendon It consequently may be partially separated in childhood by excessive strain exerted through the Achilles tendon and symptoms may develop which resemble apophysitis Apophysitis represents a true degenerative affection involving the epiphysis It occurs most often in boys between the ages of 10 and 15 years

The etiology of apophysitis is unknown It may result from (1) direct trauma or strain from the attachment of the powerful heel tendon (2) circulatory or nutritional disturbances of the epiphysis probably secondary to trauma or (3) systemic infections

The first symptom is usually a slight limp which is followed by definite pain localizing on the posterior aspect of the os calcis at the tip of the heel An area of tenderness over the tip of the heel may be elicited by the lateral palpation of the heel with the index finger and thumb Usually the line of tenderness to pressure follows the outline of the calcaneal epiphysis There is at times some swelling of the lateral and medial aspects of the heel and in cases of long standing edema may be present A roentgenogram taken in the lateral plane will show the epiphysis to be irregular and segmented with areas of increased density

Tenosynovitis of the Achilles Tendon This condition is similar to tenosynovitis involving any tendon sheath The condition is characterized by local pain swelling and redness over the Achilles tendon and crepitation over the tendon on movement

Bursitis The bursa most commonly involved is the retrocalcaneal bursa which lies between the os calcis and the Achilles tendon Inflammation of this bursa is characterized by local pain tenderness and swelling The swelling when present is localized on the sides of the Achilles tendon Local periostitis with proliferative changes and irregular spur formation on the posterior surface of the os calcis are occasionally found associated with inflammation of this bursa and may be the cause of the bursitis If the bursitis does not subside under conservative measures the bursa should be excised

Friction of the heel counter of the shoe may cause bursitis of the superficial calcaneal bursa which lies between Achilles tendon and the skin Such a superficial bursitis can usually be relieved by the application of heat and protecting the area by an adhesive strapping

AFFECTIONS OF THE NAILS

Ingrowing Toenail (Onychocryptosis—Onychia) Onychocryptosis is an acute inflammatory reaction often frank infection of the soft tissues at the corner of the nail of a toe The nail of the great toe is most often involved

On observation of the stance of the foot in individuals complaining of ingrowing toenails faulty foot balance will usually be found Pointed shoes which make pressure on the nail of the great toe careless and improper trimming of the nails and excessive width and abnormal convexity of the nail result in local irritation and may eventually cause an ingrowing nail

Pain tenderness swelling and redness at the corner and along the lateral margin of the nail characterize the affection Frequently there is frank infection of the involved area with pus formation subsequent to which the skin breaks down and a discharging area covered with sluggish granulations develops

coccus The bacteria may reach the synovial tissue by hematogenous dissemination invade the joint by direct extension from a suppurative process in adjacent bone such as osteomyelitis or they may be introduced into the joint cavity by a penetrating wound

The synovial tissue is hyperemic swollen thickened and infiltrated If suppuration continues for several weeks the result is ulceration and irreparable destruction of the articular cartilage When there is severe damage of the cartilage ankylosis results Abscesses may form in the marrow of the subchondral bone The soft periarticular structures often become acutely inflamed

SYMPTOMS The onset is often sudden with fever chills sweats malaise and anorexia The joint is tender red warm swollen and painful Its capsule is distended by effusion and the adjacent muscles are in spasm Tenosynovitis and bursitis may be present Commonly the hip knee elbow or shoulder is affected although no joint is exempt It is rare that more than one or two joints are affected In some cases the systemic disease may be so severe and the arthritis so mild that the joint infection is not noticed In others the arthritis may be unaccompanied by constitutional signs and the probable primary focus of infection remains obscure

EXAMINATION The joint or joints are red swollen and tender There is a definite leukocytosis with a rise in the percentage of polymorphonuclear cells The erythrocyte sedimentation rate is elevated Blood culture will often reveal the causative microorganism before chemotherapy The synovial fluid is cloudy or frankly purulent and the leukocytes may number more than 100 000 per cubic millimeter with more than 90 per cent polymorphonuclear cells On bacteriologic examination (both smear and culture) of aspirated synovial fluid the organism causing the arthritis can usually be demonstrated

After the tenth day long after the disease should have been diagnosed roentgenograms reveal abnormalities When treatment has not been effective roentgenographic examination is indicated for demonstration of the extent of damage to the articular cartilage and to the adjacent bone

DIAGNOSIS Recovery of the organism from synovial fluid by smear or culture proves the diagnosis Acute monarthritis of abrupt onset a red warm swollen exquisitely painful and tender joint associated with fever leukocytosis and rapid sedimentation rate is characteristic of suppurative arthritis

The prognosis as to life is generally determined by the nature of the systemic disease If correct therapy and this depends on the nature of the infection is started within the first week of infection the prognosis is better than when treatment is started later

2 ***Meningococcal Arthritis*** The incidence of articular complication during the course of epidemics of meningococcal meningitis varies from 2 to 20 per cent

There are two general types of joint symptoms associated with epidemic meningitis The first type appears during the first few days of the disease when there may be *a transient multiple synovitis which resolves spontaneously* without progressing to effusion or suppuration Normal joint function is restored in a few days

The second type a purulent usually a monarticular arthritis commences toward the end of the first week of the illness The knee or the wrist is most commonly affected any other synovial joint may however be involved

The joint on examination is acutely inflamed and distended with effusion

A diagnosis of meningococcal arthritis can be proved only by recovery of the organism from the joint fluid The general prognosis is not significantly influenced by the arthritis The bacteremia and arthritis usually respond satisfactorily to sulfonamide compounds (orally) or penicillin (intramuscularly)

3 ***Gonococcal Arthritis*** Gonococcal arthritis results from a spread of the organisms from the primary focus in the urogenital tract through the lymphatics and blood stream to the synovial tissue Arthritis usually appears within 10 to 30

The rapidity with which swelling of a joint takes place in some instances seems to determine the intensity of the symptoms. This is not borne out in all instances by clinical observation, for a knee joint affected by that strange condition known as intermittent hydrops may swell rapidly and the swelling is not accompanied by pain. Those swellings which appear rapidly and are acutely painful are likely to be associated with more intense inflammation than other swellings. It is well, therefore, to know that swelling due to edema of tissue is painless in the absence of an *inflammation of the joint structures.*

Referred pain may be felt in the joints from visceral disease, for example, shoulder pain in coronary artery disease, and, conversely, pain suggestive of visceral disease may be referred from skeletal lesions. Pain arising in one joint may be felt in another, for instance, the pain referred to the knee in arthritis of the hip.

The tendency of *emotional shocks* and *stresses* to be followed by exacerbations of symptoms may occur in those who have rheumatoid arthritis. The etiology of the arthralgias complained of by psychoneurotic patients is obscure.

POLYARTHRITIS

Various classifications of arthritis comprising an enumeration of the groups of arthropathies have been published. The groups of arthritis which usually are included are as follows: (1) infective, (2) traumatic, (3) joint diseases secondary to peripheral vascular disease, (4) joint disease secondary to blood dyscrasias, (5) allergic arthritis, (6) joint diseases of metabolic origin, (7) neuropathy secondary to disturbances of innervation or psychic control, (8) static or mechanical abnormality—loose bodies in the joint, (9) diseases of unknown or uncertain causes and (10) new growths.

Infective Arthritis. This group includes all forms of arthritis in which a bacterial or a viral etiology is demonstrated. There are two groups of infective arthritis: (1) the primary specific infection of the joint, which is characterized by bacterial invasion of the joint from without; (2) the secondary specific infection of the joint which occurs from within although the bacteria may not be demonstrated.

The primary specific infection is proved by bacteriologic study, and the bacteria are likewise demonstrated in the joint. The bacteria have been carried to the joint by the blood stream. The microorganisms which may thus cause joint disease are (1) staphylococci, (2) streptococci, (3) pneumococci, (4) meningococci, (5) gonococci, (6) tubercle bacilli, (7) *Brucella melitensis* (malta fever, brucellosis), (8) *Klebsiella granulomatis* (granuloma inguinale) and (9) *Treponema pallidum* (syphilis).

The secondary infective arthritis, in which the bacteria may or may not be demonstrated in the joint but in which arthralgic or arthritic manifestations accompany the primary disease, are: (1) scarlet fever, (2) samonellosis, (3) coccidioidal arthritis. These arthropathies are considered with description of the primary disease elsewhere in this book.

The systemic diseases in which a filtrable virus is the causative agent and in which arthralgia and joint changes are present are dengue fever, measles, mumps, chickenpox, smallpox and Reiter's disease. The joint diseases caused by the organisms or virus of dengue, measles, mumps, chickenpox and smallpox are mentioned in descriptions of these diseases. Reiter's disease is discussed in the portion of the text on infective arthritis.

The arthritides caused by the staphylococci, streptococci, pneumococci, gonococci and meningococci are often referred to collectively as *acute pyogenic arthritis*. Acute pneumococcal arthritis, though rare, may occur during the course of convalescence from pneumococcal pneumonia.

1. ***Staphylococcal and Streptococcal Arthritis.*** More than one half of the pyogenic arthritides are caused by the staphylococcus and the hemolytic strepto

expressed from a sinus It may be demonstrated by smear or recovered by culture or after inoculation of suitable material into guinea pigs

Diagnosis The diagnosis is established by demonstration of the tubercle bacilli in the synovial fluid A positive reaction to the tuberculin test is only suggestive evidence of tuberculous arthritis A negative reaction is strong evidence against the diagnosis A presumptive diagnosis can be made by the roentgenologist

5 ***Brucellosis*** *(Malta Fever)* Brucellosis is not a common disease of the bone and rarely affects the joints Approximately half of the patients who have brucellosis experience arthralgia or myalgia although only 10 per cent emphasize bone or joint pains as their chief complaint and in but a fraction of this group are there objective signs of bone involvement Permanent joint damage is rarely a result of brucellar infection (see Chapter 17)

There are no pathognomonic clinical features of the systemic infection or the joint involvement A positive culture of blood or synovial fluid is conclusive but the former is obtained in only 1 of 4 cases and the latter even more rarely An agglu tination test a complement fixation test a bactericidal test a skin test and an opsonocytophagic test have been developed but are difficult technically and of limited value because of false negative as well as false positive reactions No single one of these tests is diagnostic A positive culture from tissues is diagnostic

The duration of brucellosis is variable and may be protracted About 3 to 4 per cent of patients succumb to the infection The arthritis does not influence the prognosis significantly It is usually mild and rarely results in permanent deformity The disease is self limited and subject to spontaneous remissions

6 ***Syphilitic Arthritis*** The reported incidence of syphilitic disease of the joints decreases as the precision of diagnosis of joint disease increases

Arthritis occurs in the course of both congenital and acquired syphilis In the acquired type it may be due either to direct spirochetal infection of the joints or to neurogenic changes (Charcot s joints) secondary to tabes The latter constitutes about half of the cases in the acquired group

Generalized aching of the bones may be present during the height of the early systemic reaction in acquired syphilis

In a syphilitic infant a joint may be markedly swollen with a limitation of motion but practically without pain on manipulation (Parrot s pseudoparalysis) In children from 8 to 16 years of age a symmetric painless synovitis is the commonest form of syphilitic arthritis (Clutton s joints) Interstitial keratitis is often associated The knees are most commonly affected There is bilateral effusion without redness and with only slight pain tenderness or warmth

A monarticular gummatous arthritis may occur in late tertiary syphilis congeni- tal or acquired A gumma may arise in bone and extend into an adjacent joint or it may originate in the synovia or other soft articular or periarticular structures Swell ing with little or no pain and without stiffness local heat or muscle spasm is char acteristic Bone destruction with osteolytic areas is frequently revealed by roentgeno grams Response to antisyphilitic therapy is the rule

In those who have a Charcot joint as the result of neurosyphilis the reaction to serologic tests of blood and spinal fluid may be negative Diagnosis is based on the neurologic signs of tabes A Charcot joint also results from syringomyelia Anti syphilitic therapy is ineffective

The diagnosis is proved by positive findings on biopsy of synovial or juxta articular tissue A presumptive diagnosis is made on the presence of (1) positive serologic reaction (2) other stigmas of congenital or acquired syphilis (3) re sponse to penicillin therapy or (4) characteristic changes in the bones seen in roent genograms

Reiter s Disease Reiter s disease a secondary specific infection is due to a filtrable virus a pleuropneumonic organism This is a rare form of acute arthritis

days after the initial infection However it is said that months to years may intervene between the urethritis and the arthritis

The onset of symptoms is sudden with fever chills and joint pains The constitutional symptoms are not severe enough to direct the attention away from the joint The joint pains are those of an acute migratory polyarthritis and often it is from a few days to a week before the localization becomes monarticular

On examination in the period of the migratory joint pains, there are no definite findings except usually an active gonorrhea As soon as there is localization to one joint tenosynovitis is present Tenosynovitis occurs more frequently than arthritis particularly when the wrists or small joints of the hands or feet are affected There are leukocytosis and accelerated sedimentation rate

Cultures of exudate and synovial fluid yield positive results more frequently than do smears Occasionally however the smear is positive and the culture is negative

The reaction to the gonococcus complement fixation test becomes positive within 10 to 30 days after the onset of the urethritis in 9 of each 10 who have the disease Chemotherapy apparently does not affect the specificity of the test if arthritis has supervened This test alone does not prove the presence or the absence of gonococcal arthritis There are no characteristic roentgenologic changes in gonococcal arthritis which distinguish it from other types of suppurative joint disease

The diagnosis of gonococcal arthritis can be made only by a demonstration of the gonococcus by culture or smear of the synovial fluid Helpful diagnostic points are history of exposure and a characteristic clinical course of the disease A positive reaction to the complement fixation test and recovery or improvement with penicillin or sulfonamide therapy are only presumptive diagnostic evidence

4 ***Tuberculous Arthritis*** About 1 of each 25 who have tuberculous infections have skeletal involvement A tuberculous spondylitis is the most serious of the bone and joint infections (see tuberculous spondylitis p 281)

Tuberculosis of the peripheral joints may result either from direct bacillary invasion of the synovial tissue (tuberculous synovitis) or from penetration into the joint by tuberculous tissue from adjacent diseased bone The formation of granulation tissue separates the articular cartilage Caseation abscess and sinus formation may result

SYMPTOMS In the peripheral joints there is an effusion into the joint There are no signs of acute inflammation Articular pain and tenderness are absent until late in the course of the disease In tuberculous arthritis muscular atrophy is relatively early and prominent

The commonest complication is usually the primary pulmonary infection Abscess formation in the paravertebral region or in the vertebral bodies when the spinal column is affected and in the bone or periarticular soft structures when a peripheral joint is involved is often serious Sinuses originating from an abscess or a joint often form and when present suggest the diagnosis In Pott s disease meningitis and paraplegia may occur It seems that tuberculous bone and joint disease is a common cause of secondary amyloidosis

Tuberculous arthritis follows a protracted course of progressive destruction of bone and articular structures lasting several years Recovery of normal joint function is rare In no other type of arthritis is the mortality rate so high as in tuberculosis of the spinal column

EXAMINATION The involved joint is swollen The doughy feeling and swelling are due chiefly to edema and infiltration of the synovial tissue Redness of the joint is absent in the advanced stages unless the joint is secondarily infected following aspiration of fluid

The total and differential leukocyte counts are usually normal The erythrocyte sedimentation rate is elevated On bacteriologic study the tubercle bacillus will be found in the synovial fluid synovial tissue or the pus aspirated from an abscess or

ness Limitation of motion is due to the pain of inflamed joint structures and the associated muscle spasm A distention of the joint cavity may produce but little pain

The diagnosis of acute traumatic synovitis is usually obvious from the history the symptoms and the examination Aspiration of the joint is not routinely practiced but in some instances it may be the surest and sometimes the only way of detecting fluid in the synovial cavity and it serves the added advantage of revealing the kind of fluid present Microscopic and bacteriologic investigation of the fluid is often useful and important diagnostically

Diseases of the Joints Due to Circulatory Disturbances ***Diseases Secondary to Peripheral Vascular Disease*** *Aseptic necrosis of bone and joint* may occur secondarily to an infarct or other localized loss of arterial blood supply to bone adjacent to the joint In some instances there is an intra articular fracture or a dislocation Aseptic necrosis results in pronounced irregularity in the contour of the articular surface with secondary degenerative joint disease

Aseptic necrosis never eventuates if full restoration of the contours of the intra articular surfaces occurs it therefore differs from traumatic aseptic necrosis in that under favorable conditions restoration of normal articular contours may be expected in this disease

Joint disease and osteomyelitis occur secondarily to disturbances of blood flow during the progress of Raynaud s disease scleroderma thromboangitis obliterans (Buerger s disease) ergotism and periarteritis nodosa

Disease Secondary to Blood Dyscrasias Hemorrhage into a joint or joints of the extremities is a common manifestation of *hemophilia* The joint becomes swollen painful and stiff from intra articular bleeding The hemorrhage into the joint may be slight or so great that pain and swelling continue for weeks Once a joint has been affected it is likely to be affected again and again After repeated small hemorrhages or a large hemorrhage into a joint has occurred the joint ultimately becomes ankylosed One or more deformed ankylosed joints are present in practically every hemophiliac who has reached adulthood

Evidence of increased coagulation time of the blood in association with roentgenologic findings of atrophy of bone and periosteal thickening is sufficient for diagnosis Pseudohemophilia causes the same sort of joint symptoms

Allergic Arthritis Experimental work indicates that there are true allergic forms of arthritis such as those observed during serum sickness and possibly sequential to the administration of drugs particularly sulfonamide compounds and penicillin The opinion is abroad that sensitivity to foods may cause transient or permanent joint changes but proof is lacking True allergic arthritis aside from serum sickness is rare the diagnosis should be made with caution

Criep differentiated five types of allergic arthropathies (1) certain instances of chronic arthritis which are thought to be due to bacterial allergy (2) articular swelling resulting from sensitivity to a foreign serum or to a drug (3) certain intermittent hydrarthroses (4) some instances of Henoch s purpura associated with articular swelling and pain and (5) certain instances of acute transient paroxysmal articular involvement of joints characterized by pain swelling and limitation of motion

The mechanism of production of symptoms is probably the same as that in urticaria or angioneurotic edema the shock tissue is the synovial membrane of the joint instead of the skin

Hench has repeatedly stated in his publications that he has not been able to diagnose chronic allergic arthritis

Intermittent Hydrarthrosis Intermittent hydrarthrosis may be a manifestation of various joint diseases or of early rheumatoid arthritis The condition often affects the knees bilaterally In some instances intermittent hydrarthrosis may be allergic in origin

which is said to be limited to men It causes mild to moderate constitutional reactions In size this organism stands midway between viruses and rickettsiae

There is a triad of symptoms namely (1) acute polyarthritis (2) urethral discharge and (3) conjunctivitis The onset is acute and the arthritis the conjunctivitis or the urethral discharge may come first There may be fever

On examination one or more joints usually of the hands are red swollen and moderately tender The urethral discharge may be mild or profuse and is negative for gonococci The conjunctivae are congested and often painful Conjunctival smears and cultures are negative for gonococci The arthritis lasts for weeks or months and tends to recur

The diagnosis is made by a process of elimination The disease is self limited Rheumatoid arthritis is to be differentiated

Arthritis Due to Infection by Higher Animal Parasites Generally the joints seem to be immune from invasion by animal parasites If joints are affected from parasitic infestations it is secondarily as the result of a sensitization to a toxin elaborated by the parasite as has been suggested by Rappaport in a discussion of intestinal amebiasis

Traumatic Arthritis The term traumatic arthritis applies to various injuries in and about the joints Traumatic arthritis may be caused by trauma applied extrinsically or intrinsically to the joint It may be manifested either acutely or chronically

Acute extrinsic arthritis is often caused by a single *extrinsic* cause for instance the joint is struck a single blow which injures the intra articular surface of the joint However acute joint symptoms are manifested if the structures about the joint are so injured as to result in for example a traumatic synovitis In the absence of an *injury acute fibrositis myositis bursitis and tendinitis may occur spontaneously* and may be attributed to injury when there has been none

Often as a result of occupational or recreational repeated trauma chronic arthritis arises It is not always possible to distinguish this type from some forms of degenerative joint disease such as osteoarthritis As in acute traumatic arthritis it may be difficult to know whether a chronic joint disease is caused from chronic spontaneously occurring fibrositis myositis bursitis and tendinitis or from trauma

Intrinsic injury of a joint usually causes a chronic disability Acute traumatic synovitis however may be due to sprains strains or fractures into a joint

Chronic traumatic arthritis may be caused by repeated or continuous minor trauma such as occurs as a result of faulty body mechanics for instance postural scoliosis or flat feet This type is indistinguishable from osteoarthritis Chronic traumatic arthritis may originate from avascular necrosis or from loose bodies (joint mice)

An accumulation of fluid in the joint cavity (hydrops hydrarthrosis) is one of the first symptoms in the joint characteristic of synovitis The presence of fluid in the joint is manifested by pain swelling obliteration of the normal bony landmarks of the joint and a sense of fluctuation on palpation The same reaction may occur in a mild degree when normal joints which have been immobilized for some time are first used

In the knee the fluid lifts the patella from its normal contact with the femur an effect which can often be demonstrated by lightly punching the patella against the femur producing an audible or palpable tap

In the hip and shoulder swelling of the joint may be obscured by the overlying muscle However a comparison in size with the opposite side reveals the degree of swelling present

In all forms of acute traumatic synovitis the pain is severe The intensity and distribution of the pain are enhanced by associated lesions of bone ligament or tendon The involvement of these additional structures is detectable by local tender

ciency leukemia polycythemia vera and pernicious anemia The serum uric acid is constantly elevated in chronic gout

Therapeutic Test Colchicine if given promptly and in sufficient doses will usually relieve the acute manifestations of gouty arthritis Other forms of arthritis are unaffected by colchicine

Neuropathic Arthritis Joints atrophy when their nerve supply is badly de ranged or discontinued A neurotrophic joint disorder may be secondary to tabes dorsalis syringomyelia nerve injuries or leprosy Hysterical and psychogenic dis turbances also cause joint dysfunction

In syringomyelia and in tabes disturbances of joints are common Spontaneous fracture may occur from increased fragility In the Morvan type of syringomyelia there may be gradual disintegration of a fingertip similar to that which occurs in leprosy and in Raynaud s disease

A perforating ulcer of the leg or foot caused by tabes involves at first the soft parts later on it may cause destruction of the underlying bone

Charcot s Joint The pathogenesis of Charcot s joint is not understood How ever it is thought that this disorder is a rapidly developing degenerative process in joints lacking the protection of pain and position sense and hence used in an awk ward and traumatizing manner

The onset may be gradual or sudden and the course progressive or stationary One large joint of the lower extremities is usually affected by a painless enlargement and hypermobility Occasionally but rarely the process is multi articular and symmetric

The flail joint is enlarged and deformed and can be painlessly hyperflexed extended and rotated in several directions Warmth redness and tenderness are absent

The diagnosis is made clinically on the basis of a neurologic examination in association with history and complaint On roentgenologic examination there are usually destructive and productive bone changes with pathologic fractures disin tegration and disorganization of the joint

Psychogenic Arthropathy There are functional derangements of joints with out structural change considered to be psychogenic The hysteric joint belongs in this group Recently considerable emphasis has been placed on what some call psychogenic rheumatism Some observers have believed that these psychogenic conditions when of long standing may result in true organic changes

Psychoneurotic individuals in emotional conflict caused by psychic trauma fear anxiety apprehension or sorrow may experience arthralgia muscle and tendon aches stiffness interference with joint motion and other rheumatic symptoms which resemble arthritis or fibrositis

On examination by physical means there is absence of joint changes Roent genologic examination and laboratory observations reveal that organic health is good

In some instances if a psychoneurosis is present bizarre deformities which do not conform to anatomic changes are helpful diagnostic features It is difficult to differentiate psychogenic rheumatism from fibrositis In psychogenic rheumatism however there is a lack of consistent localization of symptoms a lack of benefit from analgesics heat and physical therapy and there is failure to improve in a warm atmosphere and after mild exercise

The Static Influences on Joints An extremity is a static unit and disturbance in one part of this unit will secondarily affect other parts This static factor is symp-tomatically important in any joint disease

In this connection two interesting changes that are met with about the joints may be mentioned although they are easily recognized the beginner may be puzzled

Suddenly without prodromes, the knees feel stiff The patient observes that the joints are swollen There may or may not be pain After varying intervals the swelling disappears to recur weeks or months later Several such attacks may occur without leaving any disability

The general condition is good Rarely are there other manifestations of the allergic state

There are no definite diagnostic criteria The knees are swollen or distended with fluid The diagnosis is made with caution It seems however that intermittent hydrarthrosis may exist as a separate disease entity

Joint Diseases of Metabolic Origin Gout and Gouty Arthritis In gout there is a concomitant disturbance of purine metabolism and the disease is spoken of as being a metabolic arthritis The definite etiology is not known The disease is characterized by (1) often a family history of gout (2) acute attacks of arthritis with freedom from joint symptoms between attacks unless chronic deforming changes have developed (3) increased concentration of uric acid in serum and other body fluids, (4) deposition of sodium urate crystals in articular periarticular and subcutaneous tissues and often uric acid stones in the urinary tract and (5) degenerative changes in the vessels of the kidneys heart and brain as the disease advances

At least 9 of every 10 patients who have gout are men more than 30 years of age The disease occurs in all races nationalities and social levels irrespective of their manner of living Excessive ingestion of purine foods or alcohol may provoke an attack of gouty arthritis in those possessing a gouty diathesis but does not cause this error in metabolism Likewise surgical operations dietary indiscretions emotional stress administration of crude liver extract thiamine dehydrocholic acid (Decholin) and mercurial diuretics may precipitate an acute attack

Symptoms of Acute Gout Within a period of a few hours the affected joint or joints become red warm swollen tender to touch and acutely painful when moved Weight bearing is often impossible when the affected joint is in a lower extremity for instance when the metatarsophalangeal joint of the great toe is affected The metatarsophalangeal joint of the great toe is affected in about half of the patients but other toes or joints of the feet ankles knees hands wrists or elbows may be involved A moderate fever leukocytosis and an increased sedimentation rate may be present The arthritis may be monarticular or migratory and polyarticular Joint synovitis with effusion and acute bursitis are often associated Specific treatment during the attack ameliorates the symptoms within a few days otherwise severe symptoms may last for several days or even weeks until the disease is far advanced After the acute attack the joint is free of symptoms until the next acute attack Finally the joints are so injured by repeated attacks that chronic joint disease consisting of swelling pain partial or complete ankylosis is present

Examination In addition to the tender red swollen joint uric acid tophi may be present They occur most often about the margin of the ears and less often in the olecranon region and around joints of the hands and feet If a tophus is suspected its contents should be removed and examined microscopically for needle like crystals or chemically by means of the murexide test

In chronic gout the joints may be found to be in various states of dilapidation these joints are the source of great agony and total disability

During the earlier stages or years of the disease the joints may appear normal on roentgenologic examination As attacks recur osseous tophi are observed as punched out regions in the bones of the fingers or more often in the bones of the feet

Diagnosis The diagnosis is based on the characteristics of the disease as enumerated

The serum uric acid is increased during the acute attacks Nongouty conditions which may be associated with an increase in serum uric acid include renal insuffi

Rheumatic Diseases lists the theories of etiology as follows (1) an infectious disease (2) a metabolic disease (3) a disease with an endocrinologic basis (4) a disease of the peripheral circulatory apparatus (5) a disease of the nervous system (6) a psychogenic disease and (7) a disease of hypersensitivity Data available now strongly suggest that rheumatoid arthritis belongs to those diseases arising from chronic anaphylaxis or hypersensitivity

Rheumatoid arthritis is a systemic disorder The disease affects primarily the interfibrillar substance of connective tissue This substance is a colloidal jelly like material existing in the interstices between cells and fibers of mesenchymal origin This substance is often termed collagen and the diseases resulting from disturbances in this substance collagen diseases

Articular lesions consist of a thickening of the synovial lining As the disease pro gresses the articular cartilage is softer contains larger ulcerations and connective tissue appears in the subchondral marrow spaces The cartilage in time may be destroyed and ankylosis may occur The cortex of the adjoining bones becomes thin and porous

The *subcutaneous nodules* of rheumatoid arthritis are a characteristic pathologic lesion of the disease Exudation of plasma and blood cellular constituents is a prominent feature of *rheumatic fever nodules* which contain in addition small focal regions re sembling Aschoff bodies In the nodules of rheumatoid arthritis proliferation and degen eration predominate Aschoff like nodules are rare and exudation is not a prominent feature The blood vessels in these nodules are affected by perivascular infiltrations ad vancing to diffuse inflammation and degeneration of the entire vessel wall Thrombosis may be present

SYMPTOMS The first symptoms often are fatigue to the point of exhaustion lassitude muscular stiffness loss of weight and general debility One or more joints slowly become swollen and painful and function is hampered The symptoms may commence with repeated mild attacks of polyarthritis accompanied by pain swell ing and redness of the affected joints slight fever and tachycardia The joints of the hands usually are affected first When the feet are affected the foot pain is most severe after inactivity The pain may be somewhat relieved by activity this is in contrast to the pain of flat feet which is relieved by rest and made worse by activity

There are some patients in whom rheumatoid arthritis commenced with an acute polyarthritis high fever and sweats The polyarthritis advances rapidly and quickly reduces the patients to semi invalidism or invalidism within a period of weeks

In almost all instances of rheumatoid arthritis the smaller joints of the hands particularly the proximal interphalangeal joints of the fingers are likely to be most severely affected

EXAMINATION A rheumatoid joint is swollen and there are pain and limitation of movement Owing to thickening of the periarticular tissues about the joint and the atrophy of the muscles proximal and distal to it the joint presents a spindle shaped appearance There is rarely much fluid in one of these joints The adjacent bursae and tendon sheaths are frequently involved and swollen

The muscles about a joint are antagonists The stronger of the antagonists the flexors exert more force on the joint therefore the joint is often found in a flexion contracture Flexion contractures are likely to be particularly apparent in the hands wrists knees elbows and toes but almost every joint in the body may be involved In cases of advanced disease fibrous and bony ankylosis may occur accompanied by subluxation or dislocation of the affected joints

Muscular weakness and atrophy are often prominent findings in rheumatoid arthritis and in some they are the characteristic features of the invalidism Atrophy in the muscles of the hands is often advanced However atrophy occurs in all the muscles of the extremities of which the joints are affected It seems that the atrophy of the muscles is an integral part of the disease and is not solely attributable to disuse and muscular malnutrition Atrophy is not limited to the muscles joints and

by them (1) lipoma arborescens and (2) ganglion (see Diseases of Tendons this chapter)

Lipoma Arborescens In obese women of about middle life it is very common to find below either the medial malleolus or the lateral malleolus a lobulated flat tumor-like mass This is the well known lipoma arborescens It is very common and may depend on fatty degeneration of a part of the synovial membrane of the joint

Joint Diseases Associated With Mechanical Abnormalities ***Loose Bodies in Joints*** Loose bodies in the joints such as those resulting from osteochondromatosis osteochondritis dissecans loose fragments from intra articular fracture fractured meniscuses or other foreign bodies often result in sufficient irritation to produce degenerative joint disease

When cartilage or bone is torn loose in a joint cavity the fragments become worn slowly so that they can move freely about inside the cavity sometimes giving rise to recurring attacks of pain swelling and disability by their mechanical interference with joint movements Loose bodies in a joint produce symptoms only when they oppose the actions of the joint The common symptom is locking of the joint which almost always occurs during flexion The pain is severe, and often time is required to extend the joint fully after locking The condition is intermittent Long intervals may supervene between attacks

The diagnosis is made from the history Often the loose bodies may be visualized on roentgenologic examination when they contain bone or calcified areas

Spondylolisthesis This lesion consists of a forward displacement of the body of one of the lumbar vertebrae on the one beneath The most commonly displaced is the fifth vertebra which slips forward over the sacrum The etiology is obscure The condition seems to be associated with increased lordosis of congenital malformation Surgical intervention may be required The diagnosis is made by the roentgenologist

Arthritis of Undetermined Etiology In this large group of chronic arthritides of undetermined etiology the joint manifestations are of primary importance and the existing systemic diseases are of interest secondarily

Classification of these forms of arthritis is not possible enumeration can be made Such enumerations were published in *Differential Diagnosis of Chronic Arthritis* by Nutrition Research Laboratories Chicago in 1942 and in *Primer on the Rheumatic Diseases* by the American Rheumatism Association in 1934 and 1942 and in the *Journal of the American Medical Association* in 1949 Reference has been made to these sources in this text

Rheumatoid Arthritis Synonyms of rheumatoid arthritis are atrophic arthritis proliferative arthritis chronic rheumatic arthritis primary progressive polyarthritis nonspecific infectious polyarthritis

Five types of rheumatoid arthritis have been defined (1) the adult type and four variants (2) the childhood type (Still s Disease) (3) Felty s syndrome (4) psoriatic arthritis and (5) rheumatoid spondylitis (Marie Strumpell arthritis) These diseases were formerly described as separate disease entities Such descriptions seem to have originated from the fact that rheumatoid arthritis varies greatly in its manifestations in different periods of life It still is not clear that rheumatoid spondylitis is rheumatoid arthritis

1 ***Rheumatoid Arthritis of the Adult Type*** Rheumatoid arthritis has a tendency to be familial the sex incidence in rheumatoid arthritis other than rheumatoid spondylitis is 3 women to 1 man The disease often commences between the ages of 25 and 50 years with the peak at 35 to 40 years

The number and the plausibility of certain current theories of etiology of rheumatoid arthritis prevent a definite classification of this disease A further uncertainty about the cause of this disease has been actuated by the fact that symptomatic improvement follows the use of adrenocortical hormones The *Primer on the*

liver and spleen in the presence of leukopenia Some of these patients have subcutaneous nodules of rheumatoid arthritis

4 **Psoriatic Arthritis** (*Rheumatoid Arthritis*) About 3 per cent of patients who have psoriasis have rheumatoid arthritis This may be only an average tendency for these two diseases to coexist However there are those who have psoriasis in whom the arthritic changes occur only in the terminal phalanges of fingers which show overlying psoriatic lesions of skin and nails Such may be a distinct form of arthritis but at present the question cannot be definitely answered

5 **Rheumatoid Spondylitis** (*Marie Strumpell Arthritis*) Synonyms are spondylitis deformans spondylitis ossificans ligamentosa or rheumatoid arthritis of the spinal column ankylosing spondylitis von Bechterew s arthritis spondylitis ankylopoietica and spondylitis rhizomelica

The view formerly held that rheumatoid spondylitis was a separate disease entity from rheumatoid arthritis was based on the characteristic course of rheumatoid spondylitis and difference in response to therapeutic trials

The etiology of the condition is unknown Presumably it is a form of rheumatoid arthritis often originating in the sacro iliac articulations accompanied or followed by inflammation of the apophyseal joints or vertebral facets It is a disease of young adults and commences between the ages of 20 and 25 years It is 10 times more frequent in men than in women

The manifestations of spondylitis have been arbitrarily divided into three phases (1) the prespondylitic or early stage which is characterized by shifting pains of a rheumatic nature in the limbs thorax and peripheral joints (2) the sacro iliitis phase during which the patient may be able to carry on his daily work because the discomfort is less marked during the day than at night several hours after retiring and (3) as the disease progresses the poker back period which is characterized by pain along the entire spinal column stiffness and eventual rigidity Rigidity of the spine is not painful though the hip and leg joints are more susceptible to fatigue in this condition

Examination reveals a stiff back with decreased or absent expansion of the thorax during inspiration The degree of disability is largely proportional to the deformity of the spinal column The spinal column may be bent and ankylosed so far forward that it is difficult for the patient to walk because the head and eyes are directed downward toward the anterior thoracic wall Lateral deformities of the spinal column may be present a condition which adds to the disability This type of ankylosis of the spinal column illustrates well the influence of deformities of the axial skeleton on locomotion

Roentgenologic examination during the prespondylitic stage is not necessary for diagnosis The roentgenogram may be recorded as being negative The diagnosis can be made from the foregoing history and examination During the phase of sacro iliitis the roentgenologist may observe definite osseous changes which involve the sacro iliac articulations As the disease progresses changes detectable on roentgenologic examination gradually become apparent around the apophyseal joints or articulating facets the joint spaces appearing narrowed or obliterated Decalcification of the vertebrae and still later calcification of the ligaments is reported by the roentgenologist Gradually the entire spinal column roentgenologically may appear to be fitted with a calcified casing partially obliterating the individual bony structures

The condition may be confused most frequently with osteoarthritis

Once the spinal column becomes fixed the disease has about run its course The fixation is permanent

Rheumatic Fever The polyarthritis of rheumatic fever is one of the common and important forms of joint disease to be recognized particularly in regard to its association with carditis for in adults the joint manifestations may overshadow for

bones for it often affects the skin The skin over the crippled extremities is smooth glossy and atrophic The hands and feet may be cold and dry or cold and wet

Subcutaneous nodules occur in some patients and are often situated over bony prominences especially in the region of the elbows and over the ulna just distal to the olecranon They vary in size from those just large enough to be palpable to lesions 3 or 4 cm in diameter They are not painful They may become ulcerated and infected The rheumatoid nodule may persist for months to years

The spleen may be found enlarged A generalized lymph node enlargement of moderate degree is commonly observed Iritis and uveitis often intractable, may accompany or precede rheumatoid arthritis

There is often a hypochromic microcytic anemia of the iron deficiency type which responds little if at all to administration of iron The leukocyte count the differential count and the platelets are within normal limits The erythrocyte sedimentation rate is almost always rapid although a few exceptions of active disease with a normal rate may be seen Hench pays close attention to the trend of the sedimentation rate for he believes it indicates the trend of the disease The roentgenologic appearance of joints varies in different stages of rheumatoid arthritis

Diagnosis The appearance and distribution of the joint lesions in those less than 40 years of age which have been described are diagnostic The diagnosis is confirmed by the laboratory and roentgenologic findings At times in those more than 40 years of age an extensive survey has to be made in order to differentiate rheumatoid arthritis from degenerative joint disease such as osteoarthritis

Differential Diagnosis Rheumatoid arthritis is to be differentiated from degenerative joint disease or osteoarthritis Rheumatoid arthritis is a systemic disease and the patients are sick Osteoarthritis as a manifestation of aging affects hyaline cartilage and the patients are otherwise well Muscular weakness and atrophy and cutaneous changes are commonly associated with rheumatoid arthritis rarely with osteoarthritis or degenerative joint disease Weight bearing joints are more often involved in degenerative joint disease whereas in rheumatoid arthritis the involvement is usually symmetric and generalized The proximal interphalangeal joints are involved in rheumatoid arthritis the terminal most often in degenerative joint disease (Heberden s nodes) although occasionally the proximal joints are involved also In degenerative joint disease subcutaneous nodules are not present and the erythrocyte sedimentation rate is elevated only slightly if at all Elevation of this value is the rule in rheumatoid arthritis (*Primer on the Rheumatic Diseases* and in *J A M A* 1949)

There is a chronic type of rheumatic polyarthritis very similar to that of rheumatoid arthritis and differentiable only by a period of observation In chronic gout tophi an increased serum uric acid concentration and roentgenographic changes serve to differentiate the two Psychogenic joint disease may be differentiated in some instances only after a period of time

Prognosis Some patients who have rheumatoid arthritis recover completely Often the disease spontaneously becomes arrested or quiescent at some stage in its course and the patient is able to carry on activities with limited disabilities Finally there are those whose disease advances rapidly to complete invalidism of a most miserable sort It is impossible to forecast the turn of events in an individual patient

2 ***Juvenile Rheumatoid Arthritis*** (*Still s Disease*) The term Still s disease is employed now to designate the disease of rheumatoid arthritis as it occurs in children Children who have Still s disease have been observed to carry the disease into adulthood

In Still s disease the joints have the same appearance as in adult rheumatoid arthritis and there may be subcutaneous nodules

3 ***Felty s Syndrome*** (*Rheumatoid Arthritis*) The term Felty s syndrome is employed to designate rheumatoid arthritis which is accompanied by an enlarged

faces Subsequently the irregularities increase and the cartilage is more deeply affected The cartilaginous surface may show wide erosions and splits which penetrate to the epiphyseal bone Macroscopic fragments of cartilage may be separated and dislodged into the articular space There is active growth at the periphery of the joint membranes where cartilage and synovial intima form a zone of transitional tissue Marginal proliferations arise at the juncture of periosteum and fibrous synovial capsule and from subchondral bone which produce the bony spurs lipping or osteophytes These changes represent an advanced phase of osteoarthritis True ankyloses do not occur although calcification and osseous bridges are found in some joints and in interspinous ligaments leading to variable degrees of rigidity of the vertebral column

SYMPTOMS The disease commences with mild stiffness and aching pain in or about the affected joint Constitutional manifestations are lacking

Despite the steadily progressive anatomic changes which have been demonstrated in most persons more than 60 years of age the frequency of symptomatic illness from this process is not great even in older patients Pain from an osteoarthritic joint is often referred to distant parts for instance referred pain from vertebral joints may pass around to the abdomen

EXAMINATION On examination passive or active motion especially in the extremes of range may produce pain Palpation of the joint during motion may elicit crepitus Tenderness may be present at the articular margins There may be spasm and tenderness of the associated muscles As the disease progresses there is gradual firm or hard irregular enlargement of the affected joints In the terminal interphalangeal articulations there may be tenderness and soft swelling Generally articulations which are subject to greater strain such as the knee and hip joints and the lower spinal articulations show the greater disturbance of function The terminal interphalangeal joints show greater changes in size and function than the proximal interphalangeal joints The localization of the disease however affords no absolute diagnostic criterion There are no laboratory findings indicative of osteoarthritis

DIAGNOSIS A progressive but often intermittent articular aching relieved on rest with minimal physical signs of joint disease in a middle aged person without evidence of general illness suggests the diagnosis of osteoarthritis The diagnosis of advanced osteoarthritis or degenerative joint disease is distinctive when it affects the smaller finger joints of the hands with firm or hard knobby enlargement leading eventually to ugly deformities and limited range of motion

The phenomenon of referred sensations and pain from the thoracic and lumbosacral segments to the trunk and lower extremities in those who have osteoarthritis is common Neuralgias localized to the occipital frontal retrobulbar or other regions of the head may have their origin in foci of cervical spondylosis

The course of osteoarthritis is slowly and steadily progressive over years without severe acute attacks of joint redness and swelling unless the joint or joints are traumatized After acute trauma it may be for a while difficult to determine whether the patient has osteoarthritis or rheumatoid arthritis except by roentgenologic examination

NONARTICULAR RHEUMATISM

Often the term nonarticular rheumatism is employed to designate a rheumatic disease that does not affect the joints and that is associated with one or another of a large variety of illnesses Nonarticular rheumatism is poorly defined clinically and pathologically so that there is lack of agreement about the prevalence of this protean rheumatic ailment and the importance of differentiating it from arthritis

Among the diseases associated with nonarticular rheumatism should not be included the many febrile illnesses particularly infectious diseases of specific causation which in prodromal or early stages are frequently accompanied by rheumatic symptoms which may constitute the patient s chief complaint As these ailments progress their nature becomes evident so that differentiation from primarily rheu

a long time those referable to the heart (see Chronic Disease and Allergy, Chapter 18)

Rheumatic fever either in children or in adults is characterized by (1) migratory polyarticular joint involvement (2) the presence of carditis subcutaneous nodules pleuritis, erythema marginatum and chorea and (3) a quick response to the anti rheumatic action of salicylates The joint involvement is acute and is accompanied by a temperature of 101 to 104 F (38 3 to 40 C) leukocytosis and rapid erythrocyte sedimentation

The diagnosis can be made on the clinical findings

Palindromic Rheumatism Palindromic rheumatism is a rare disease charac terized (according to Hench) by multiple afebrile attacks of acute arthritis and periarthritis sometimes also para arthritis with pain swelling redness and disa bility generally of only one but sometimes of more than one small or large joint of adults of either sex The attacks appear suddenly develop rapidly generally last only a few hours or days then disappear completely but recur repeatedly at short or long irregularly spaced intervals Despite the frequent recurrences and the transitory presence of an acute or subacute inflammatory polymorphonuclear exudate in articular tissues and cavity there is little or no constitutional reaction or abnormality in laboratory tests and no significant functional pathologic or roent genographic residues occur even after years of disease and scores of attacks

The chief points which distinguish palindromic rheumatism from rheumatoid arthritis are the tendency for only one or two joints to be involved in an attack the frequent isolated short attacks of para arthritis the absence of significant constitu tional reactions and the persistently negative roentgenograms The greatest and most convincing difference is the persistent absence of chronic arthritis even after many attacks extending through years of the disease There appears to be little or no tendency for the disease to become chronic in a given joint

Osteoarthritis (*Degenerative Joint Disease Hypertrophic Arthritis*) The fol lowing are several varieties of osteoarthritis each representing different manifesta tions of the same disease process (1) Heberden s nodes (affection of terminal phalangeal joints (2) malum coxae senilis (morbus coxae senilis affection of hip) (3) osteoarthritic spondylitis (spondylosis affection of spinal column) Osteoar thritis may be further subdivided according to the extent of the involvement as follows (4) generalized—characterized by hypertrophic changes principally in the terminal phalangeal joints of the fingers in the knees and vertebrae although other joints may be affected the pathologic changes produced in the articular structures by the wear and tear of daily activity which are apparent in most people after the age of 50 years are included in this group (5) localized—one or two joints are affected which have previously been injured by (a) trauma—secondary osteo arthritis following fractures or injuries (b) infection (c) structural abnormality (congenital and acquired deformities acting as stresses) (d) poor posture and (e) muscular imbalance

The causation of osteoarthritis is unknown The primary change is one of de generation which is correlated with both advancing age and functional stresses such as weight bearing Deterioration of cartilage in the weight bearing areas for instance of the knee joint may be demonstrated in some persons as they approach 20 years of age Osteoarthritis occurs with great frequency and in many joints between the ages of 20 and 40 years and is present in most persons past 60 years of age Con tributory factors are occupational stresses obesity malposture and habit spasm Wear and tear are also increased by functional derangements or mechanical defects within a joint The clinical manifestations are seen more often in women than in men Heberden s nodes occur more frequently in women and a sex linked hereditary predisposition for this classic feature of osteoarthritis seems to be definite

Early pathologic manifestations are small pits and irregularities of the articular sur

examination may be diagnostic Fragments of the tendon are one source of joint mice

Chondrodystrophia Foetalis This disease often is erroneously termed osteo genesis imperfecta and achondroplasia Chondrodystrophia implies a nutritional disorder of the cartilages in fetal life which seems to be true Achondroplasia signi fies an entire absence of the normal activity of the cartilage in producing osseous tissue which in fetal rickets is not true

Chondrodystrophia foetalis is a rare disease of the skeleton beginning in fetal life and affecting those bones preformed in cartilage Heredity is fully established as an etiologic factor The characteristic features are present at birth a moderately enlarged head depression of the root of the nose trident hands and remarkably short and curved extremities (micromelia)

Nearly all of these dwarfs die between the seventh and ninth months of intra uterine life Of those living at birth the majority succumb within the first few months The few who survive the first year or two develop without impairment of their general health and apparently have the same life expectancy as do normal persons If the chest is well formed since the disease is primarily one of deficient bone growth after the age of maturity a normal life is expected Many have lived to extreme old age These survivors are termed chondrodystrophic dwarfs Dwarfs of this type often are featured in sideshows of circuses and carnivals

The adult chondrodystrophic dwarf (achondroplast) presents the same general characteristics as are seen in the fetus There is an abnormal development of the skeletal muscles and short bones which gives a strength relatively much greater than that of the normal man The posture is erect except in the lumbar region where there is marked lordosis due to the forward tilting of the sacrum The face appears small because of the enlarged calvarium The nose and the nasal region are flattened and there is a retraction at the root of the nose The end of the nose is hypertrophied and rounded The extremities like the nose are symmetrically and equally involved by a shortening in the proximal or root segment (rhizomelia) The arms are so much shortened that the fingertips barely touch the crest of the ilium Complete extension of the elbow joint is impossible and motion in the shoulder articulation is limited The hands are diminutive short broad and pudgy The fingers are of about equal length taper and distal to the second joint they are separated from each other like the spokes of a wheel hence the name trident hand (*main en trident*) The legs too are short and the femur and tibia are bowed Mal position of the knee joint and relaxation of the joints of the toes are common The sexual organs often are overdeveloped An enhanced sexual appetite is common to both sexes

The principal diagnostic features of chondrodystrophia are the fetal origin macrocephalic and brachycephalic head depressed root of nose and prognathous jaws normal trunk stunted growth of the extremities with resulting decentralization of the midpoint of the body bone deformities lordosis trident hand general excess of subcutaneous fat with thickened loose skin protuberant abdomen normal men tality and the roentgenologic findings

Ochronosis Ochronosis is an indefinite disease entity characterized by darken ing of the cartilages of the ears nose ribs and in later life of the intervertebral disks It is a rare disease

Three groups of cases are distinguished respectively by the presence of (1) alkaptonuria (2) staining of cartilages and tissues from the prolonged use of carbolic acid and (3) staining of the cartilages from melanin which is a derivative of proteins

In the presence of progressive destruction of the cartilage there are the symptoms of a destructive joint disease such as occur in osteoarthritis

Transmitted light will help to detect the contrast in color for instance in the

matic diseases is usually not difficult Varying degrees of nonarticular fibrous tissue abnormalities are usually associated with different forms of arthritis this secondary fibrositis contributes significantly to the patient s discomfort and disability It is important however to appreciate that there are many forms of nonarticular rheuma tism which occur entirely independently—primary nonarticular rheumatic diseases This group of ailments includes fibrositis generalized fibrositis localized lumbago painful stiff neck bursitis periarthritis (para arthritis) tendinitis (tendon attach ment syndrome) tenosynovitis fascitis panniculitis herniated subcutaneous fat herniated muscle shoulder hand syndrome and psychogenic rheumatism, for in stance rheumatic manifestations of psychoneurosis

PRIMARY NEOPLASMS OF THE JOINTS

Benign Tumors The benign tumors of the joints comprise cysts, xanthomas hemangiomas giant cell tumors and synoviomas These benign tumors are usually not diagnosable by clinical means Roentgenologic examination may or may not be conclusive The diagnosis is made from biopsy of the tissue and histologic ex amination

Malignant Tumors Malignant growths of joints occur with only moderate fre quency and may arise from any of the articular tissues Joints may also be involved secondarily by neoplastic processes of the adjacent bone or remote parts of the body These metastatic lesions are especially important when they involve the spinal column and must be kept in mind when back pain is extremely severe

The diagnosis is established by the history the physical and roentgenologic examinations and histologic study of biopsy tissue of the tumor Sarcoidosis though not a malignant tumor may produce changes and arthralgias resembling those of malignant tumors of the joints

DISEASES OF CARTILAGE LIGAMENTS TENDONS BURSAE AND FASCIA

CARTILAGE

Injury to Cartilage At the time of the acute manifestations of joint injury it may be impossible to detect evidence of trauma to cartilage However the cartilage of a joint may be injured without fracture and perhaps without tear of the joint capsule When a fracture extends into a joint the joint cartilage is always injured In such an instance the fluid in the joint will be bloody and will be demonstrable by aspiration

Semilunar Cartilage Injury to the semilunar cartilage of the knee may consist of a dislocation or actual rupture or tearing of it The internal semilunar cartilage is most commonly injured by a severe twisting and hyperextension of the knee De pending on the circumstances and the degree of an injury it may consist of a detach ment tear or crush

If the cartilage is only displaced on examination complete extension of the knee may be impossible and there are severe pain and local tenderness Locking of the knee with inability to extend the joint is a frequent manifestation Occasionally con siderable fluid accumulates in the joint Replacement of the cartilage often occurs spontaneously or may sometimes be achieved by cautious flexion and rotation of the tibia If replacement is successful it is indicated by the ability to extend the leg completely

The diagnosis may be established by the forementioned signs when they are acute However the condition may not be suspected for a long time If not and if there are changes in the density of the cartilage the findings on roentgenologic

Sacro Iliac and Lumbosacral Sprain Back pain and often serious disability attend sacro iliac and lumbosacral sprains Local muscle spasm tenderness and pain on movement are the important manifestations These disabilities may develop after a slight or nonapparent trauma or exertion Injury to ligaments of the lumbo sacral and sacro iliac joints is difficult to demonstrate Injuries of this type moreover because of the variety of lesions produced and the frequency of malingering among workmen who have sustained injuries of the lower part of the trunk represent an especially difficult diagnosis Sacro iliac and lumbosacral sprains are to be distin guished from coccygodynia and various types of synovitis neuritis sciatica and bursitis in the region of the back and hip

TENDONS

Many tendons move in sheaths which are structurally similar to joints and bursae They are frequently involved with acute and chronic inflammations when the nearby joint is thus affected In general however tendons are not frequent sites of primary serious disease Acute infections wounds and tumors of tendons occur

Prenatal influences as a cause of disease of the tendons may be manifested by a stenosis of tendon sheaths for instance a trigger finger or a snapping finger is often of congenital origin

Infections of Tendons (Tenosynovitis) Tenosynovitis is characterized by a spindle shaped swelling of the tendon sheaths which often move with the tendons On movement there is pain The skin may be reddened over the inflamed sheath On palpation of such a spindle shaped swelling grating may be felt on movements of the tendon

In tenosynovitis of the palm of the hand or of the wrist if accompanied by a purulent exudate there often is a severe inflammation A stiff deformed hand is too often the sequela of the infection

The diagnosis can rarely be made of bursitis or tenosynovitis as a separate entity since these conditions so frequently accompany joint disease Occasionally as the result of trauma tenosynovitis of the tendons about the wrist can be diagnosed

Stenosing Tenovaginitis Stenosing tenovaginitis of the extensor and abductor tendons of the thumb (De Quervain s disease) is a rather common condition The disorder may produce considerable disability due to pain in the thumb wrist and forearm It is commonest in middle aged women who use the wrist and thumb excessively Chronic trauma is the most frequent exciting cause The pain at first dull localizing in the region of the radial styloid and extending into the thumb and hand and up into the forearm is often neuralgic in character and may be worse at night than in the daytime It is aggravated by motions of the thumb and wrist In some cases the syndrome is bilateral There is swelling over the tendon sheath and localized tenderness over the tip of the radial styloid process A circumscribed thickening of the tendon sheath may be found on palpation and frequently a cartilaginous thickening is felt over the styloid process situated under the skin not adherent to underlying bone and moving with flexion and extension of the thumb The pathognomonic sign may be noted on grasping and quickly abducting the thumb ulnarward The pain over the styloid tip is excruciating Roentgenologic studies do not reveal abnormalities

Tuberculous Tenosynovitis Adams and his co workers analyzed the diagnostic and therapeutic features in a number of instances of tuberculous tenosynovitis The youngest patient complaining of symptoms that had lasted a year and a half was 15 years of age the oldest patient was 75 years of age and had had symptoms for 3 years when first seen The influence of use on incidence was shown by the high involvement of the right hand

Tuberculosis elsewhere in the body and trauma seem to be predisposing causes

cartilage of the ears The discoloration of the cartilage and the presence of homogentisic acid in the urine are diagnostically helpful A biopsy of the skin may be of value

Osteochondromatosis Osteochondromatosis is a condition in which cartilaginous and osteocartilaginous bodies are formed within and by the synovial membranes of joints and occasionally bursae and tendon sheaths

The formation of these cartilaginous and osteocartilaginous bodies follows the same stages that occur in the embryonic formation of cartilage As the bodies increase in size they may be forced to the surface to become pedunculated When the pedicles break as they frequently do the bodies become avascular and the bone in them dies The cartilage cells near the surface live nourished by the synovial fluid The loose bodies are white and round or oval with a smooth or faceted surface In number they vary from a few to hundreds

Osteochondromatosis occurs more frequently in males than in females and is usually monarticular The knee joint is involved by far the most frequently with the elbow second Other joints affected relatively rarely are hip shoulder wrist ankle temporomandibular metacarpophalangeal and interphalangeal Bursae and tendon sheaths are seldom affected

SYMPTOMS The symptoms are weakness aching and progressive limitation of motion of the joint In due time catching or locking of the joint may occur although it is usually of a transitory nature followed by slight effusion pain and stiffness There is rarely spontaneous pain except with an episode of locking The minority of patients mention feeling movable masses

EXAMINATION The joint may be normal to examination Loose or fixed bodies may be felt The joint capsule is thickened somewhat with moderate effusion present The motion of the joint may be decreased although usually not markedly

DIAGNOSIS In many instances in which large numbers of bodies are present they are demonstrable by roentgenogram Rice bodies (corpora oryzoidea) associated with chronic infections such as tuberculosis are differentiated with ease microscopically and relatively easily on the basis of history and findings When there are only a few bodies present the differential diagnosis may become difficult because loose bodies may be produced in osteoarthritis in osteochondritis dissecans and occasionally after trauma When the bodies are not radiopaque the conditions in the knee joint must be differentiated from a torn meniscus In the latter condition the symptoms are relatively more severe particularly early in the course of the condition

LIGAMENTS

Sprain (Rupture of Ligaments) Trauma often ruptures the supporting structures of a joint but does not involve the joint itself Such injuries are designated as sprains Sprains are to be suspected in every severe injury about a joint especially if there is exquisite tenderness over the ligaments and not over the bone A severe pain which develops on certain motions only is suggestive of a sprain if a fracture is not present

Sprain of the Ankle It has been shown that in many sprains of the ankle there is a tearing off of small fragments of the bone The term sprain fracture is applied to such injuries

Sprains of the ankle are often the consequence of a lateral displacement The resultant injury is frequently in the subastragalus and sometimes in the adjacent tarsal joints This condition is suspected when the pain and swelling are located below and in front of the ankle rather than around the ankle A sprain however is most frequently the result of inversion rather than of eversion of the foot In eversion because the plantar ligaments are strong the force is transmitted directly to the ankle and leg bones and thus the ligamentum deltoideum is ruptured A Pott s fracture often accompanies this sprain

tendons or muscles which lie over them to glide smoothly against underlying bone or other structures In superficial locations the bursae protect underlying structures from trauma inflicted over them Bursae likewise serve to facilitate skeletal movement

Some bursae occur naturally Bursae may form however in response to special needs when certain movements are repeatedly used Some bursae particularly those about the knee are continuous with the joint cavity and they are similar to joints in their reaction to injury

Bursae are rarely the sites of acute pyogenic infection except after injury and infection The commonest lesions of bursae follow trauma which is sometimes direct and acute but more often is the result of repeated slight injuries as observed as the cause of prepatellar bursitis

Of the almost 150 bursae in the body all those of clinical importance are situated near the major joints

Acute Traumatic Bursitis An acute traumatic bursitis may follow direct blows but more often it results from the same type of forceful movement which gives rise to ruptured tendons or ligaments

There are so many bursae in the body that the symptoms of acute inflammation and injury cannot be described for each of them Indeed such a description is not necessary for the symptoms are the same except for their situation depending on the bursa involved

Often acute traumatic bursitis develops rapidly after a relatively slight injury provided the bursa has been previously injured by repeated insults or irritations The symptoms are local pain and swelling The pain is severe if aggravated by certain motions The swelling is apparent only in the superficial bursae

There is often point tenderness over the bursa The disability frequently is severe on account of the pain limitation of motion and muscle spasm Occasionally infection develops in the bursa and progresses to suppuration

Chronic Bursitis Chronic bursitis may follow acute bursitis but frequently it develops insidiously

The pathologic changes in chronic bursitis are similar to those in the joint since chronic bursitis is often associated with chronic disease of the adjacent joint Fluid is present and villous overgrowth of the bursal lining occurs In addition however calcification of the wall of the bursa is frequent and often is revealed in a roentgenogram

The symptoms are pain often severe brought about or aggravated by certain movements which tend to compress or irritate the bursa

Tenderness is often localized to the region of the bursa Swelling may develop because of effusion which is always present and may be demonstrated by aspiration

The diagnosis can be made on the basis of the situation and localization of symptoms if the examiner is aware that a bursa is situated in that particular location When calcification is present the diagnosis is made on the basis of roentgenologic examination Excision of the bursa is often indicated and is frequently curative especially when the bursa is calcified

FASCIA

Dupuytren s Contracture This is probably an hereditary disorder which does not become manifest until middle age It is a contracture of the palmar fascia not of the tendons and the ulnar side of the palm which causes flexion deformity of the fifth and the fourth fingers There is no pain or tenderness

Herniated Subcutaneous Fat Herniation of subcutaneous fat through a hole in the fascia causes a localized painful condition The tender herniated fat may be palpated Herniated fat opportunely situated may cause a segmental nerve distribution of pain The small herniations of fat may occur along the sides and lower part of the back

in some instances Other tendons may be involved but the tendons of the wrist are by far the most commonly affected

The patient complains of a gradually developing painless or slightly tender mass on the volar aspect of the hand There is stiffness with inability to flex or extend the fingers completely and diminished strength on grasping The temperature may rise to 99 6 or 100 6 F (37 5 or 38 C)

On examination there is a spindle shaped swelling along the course of the tendon Finger motion in the later stages of the disease may cause creaking or grating sounds because of degenerated fibrinous deposits within the tendon sheath—so called rice bodies Joint stiffness and a fluctuant mass often indicate osseous disease which may be proved by roentgenologic demonstration of destruction of the carpus The diagnosis is proved by biopsy and by inoculation of guinea pigs

Tuberculosis of the tendon sheaths should receive the same systemic treatment accorded to tuberculosis of the lungs or spinal column Surgical treatment without knowledge of how the lesion is progressing elsewhere in the body involves the likeli hood of operating in the presence of advancing or exudative disease

Traumatic Tenosynovitis Traumatic tenosynovitis is rarely serious but it is a common injury of tendons It produces an inflammation of the sheath with out pouring of an excess amount of the lubricating fluid

On examination there is local tenderness along the course of the sheath and use of the tendon produces pain On motion of the tendon creaking or crepitation may be audible Later fibrous tissue may replace the fluid and fibrin thereby resulting in limitation of motion

A dislocation of a tendon may occur so that the tendon leaves the groove it normally occupies and finds a new abnormal site The tendons commonly involved are the long head of the biceps the peroneal tendons and the posterior tibial tendon Treatment consists of operative replacement of the tendon and rest for a week or two until movements without pain are possible

Painful Tendons Tendons may become inflamed although more often they are painful without evidence of inflammation accounting for the symptom complex known as tendon attachment pain This may result from trauma or from more than customary use with or without regional bursitis This is a common difficulty known as tennis elbow at the attachment of the conjoined tendon onto the humeral epicondyle

Ganglion Ganglion is a term applied to a smooth rounded mass (0 5 to 1 5 cm in diameter) which commonly presents itself in the distal portion of the wrist on the dorsal surface and may be made more prominent by flexion of the wrist It also occurs on the ventral surface of the wrist and in some other joints particularly the knee It is found in women much more frequently than in men

Ledderhose has shown that the ganglion originates from the fibrous tissue of the joint capsule as a neoplasm The cystic nature of the tumor is explained by the presence of secreting mesothelial cells or by mucoid degeneration of the fibrous tissue The lumen of the cyst is commonly a closed cavity but occasionally it communicates with the wrist joint or a tendon sheath

The first symptom is the presence of the tumor Later there is pain or discomfort which may result in definite disability

The round cystic tumor attached to a tendon is usually sufficient for diagnosis Breaking the cyst with a well directed blow such as that obtained from the flat sur face of a book is sometimes followed by cure but is not recommended The tumor if producing disabling symptoms should be surgically excised

BURSAE

Bursae are related to joints in that they are potential cavities lined by modified fibrous tissue or mesothelium which secretes a mucoid lubricant and thus enables

improvement of symptoms by the use of heat and massage and by the administration of acetylsalicylic acid are the characteristic symptoms of primary fibrositis These same characteristics are observed in rheumatoid arthritis

The differential diagnosis of periarticular fibrositis and rheumatoid arthritis is based on evidence of intra articular disease (such as an increase in synovial fluid) synovial swelling and roentgenographic signs of the disease Indirect evidence comprises capsular swelling redness and tenderness loss of weight anemia rapid sedimentation rate of erythrocytes muscular atrophy and limitation of the range of movement of the joints During the beginnings of either disease the diagnosis may have to wait until the manifestations are definite

A patient who has *psychosomatic rheumatism* has pain or stiffness which is more adequately described as a sensation of fatigue and weakness than as actual discomfort The patient feels that movement of an arm or leg is impossible but when he does move it the act of movement does not increase the discomfort Pains in psychosomatic rheumatism rapidly migrate beyond all bounds of a nerve and muscle group distribution They go up an arm down through the body and out through a leg and foot

Fatigue especially nervous fatigue accentuates the symptoms of psychosomatic rheumatism or primary fibrositis Mental stimulation resulting from a party will relieve the pain of psychosomatic rheumatism for hours This is not so in the case of primary fibrositis

Acute or subacute primary fibrositis usually lasts only a few days or a week or two and disappears without any apparent residua Chronic primary fibrositis may persist for years but the patients usually are able to continue their work and customary activities

DISEASES OF THE BONES

It is not always possible to determine whether a disease of bone originated in the affected bone itself in the structures of an adjacent joint or as the result of some systemic disease For this reason uncertainties in regard to classification of some bone diseases are inevitable

The age of the patient may be helpful in the identification of the nature of a bone disease for instance acute osteomyelitis is common in children but is rare in adults Separation of the epiphyses occurs only in children As age advances however the bones grow more fragile and an old person may fracture the neck of the femur by such slight trauma as catching a toe in the bedclothes

When the shafts of the long bones are soft and become curved deformities arise (for example bowlegs) It may be possible to demonstrate an abnormal softness by producing a passive curvature If the bones are spontaneously bent near the joints as in genu valgum the deformity may be less noticeable Deformities of the feet (talipes) of the hands and of the spinal column make up a large part of common deformities of bones

The common deformities in the spinal column are curvatures with a convexity backward (kyphosis) forward (lordosis) or to the side (scoliosis) Distinction is made between the deformities of curvatures and the angular deformity (gibbus) so characteristic of caries of the vertebrae and those resulting from some fractures

In the long bones of the extremities curvature arising from softening of the whole bone is most often due to rickets or osteomalacia whereas angular deformities are due to local affections of the bone such as those resulting from pyogenic organisms tuberculosis brucellosis Paget s disease hemangioma metastatic malignant lesions and syringomyelia

Asymmetric shortenings of the extremities may be due to nervous lesions or to injuries to the epiphyseal cartilage during growth Shortening of one leg causes tilt

Fibrositis Synonyms for fibrositis are myofibrositis muscular rheumatism and myositis The disease occurs in primary and secondary forms The *primary* form of fibrositis is a nonsuppurative acute subacute, or chronic inflammation of fibrous tissue without demonstrable systemic disease It may involve fibrous tissue in any part of the body *Secondary fibrositis* is a nonsuppurative inflammation of fibrous tissue that is a manifestation of a systemic disease or is due to a local injury or strain It may be a prominent manifestation of such systemic diseases as rheumatoid arthritis osteoarthritis rheumatic fever gout or lupus erythematosus It also may be due to gonococcic streptococcic staphylococcic or meningococcic septicemia or to the toxemia caused by such diseases as typhoid fever dengue or influenza Traumatic fibrositis is secondary to an acute injury It also may develop insidiously as a result of strains of fascia tendons or joints owing to poor posture as observed in scoliosis and flat feet

The cause of fibrositis is unknown No changes have been demonstrated in synovial fluid or in the intra articular membranes

SYMPTOMS The symptoms in primary fibrositis may be acute or subacute Often there are muscle pains and mild discomfort However there may be recurrent attacks of varying severity which last only a few days or weeks The attack disappears without residua but will recur After several attacks chronic fibrositis may ensue Seldom does primary fibrositis start insidiously A characteristic of primary fibrositis is its variability and its tendency to shift to other regions Only occasionally in an acute attack will there be a slight rise of temperature and minimal local swelling A prominent manifestation is subjective stiffness of the affected areas with the very first movements as soon as the patient awakens in the morning After moderate exercise the affected regions feel better Later in the day however the aching and subjective stiffness may return and get worse The affected regions seem to jell with inactivity and improve with moderate exercise The subjective stiffness and aching are often affected by changes in the weather exposure to a draft or air conditioning Acetylsalicylic acid and local application of heat afford relief

The *localized forms* of fibrositis give symptoms peculiar to the topographic and anatomic situations as follows

Periarticular fibrositis most frequently is the result of trauma rheumatic fever rheumatoid arthritis gout gonorrhea or periarthritis It causes local regions of tenderness and may produce swelling of the side of the capsule of the joint If the disease is acute the periarticular tissues may be acutely inflamed and painful Any movement of the joint may be so painful that it cannot be tolerated If the disease is chronic inactivity causes subjective stiffness but this is relieved by moderate exercise

Tendinous and fascial fibrositis most frequently is associated with Dupuytren like contractions injury of the supraspinatus tendons gout gonorrhea or rheumatoid arthritis that affects the tendons especially the tendo achillis It causes localized pain and tenderness especially when the affected tendon or fascia is stretched

Perineural fibrositis is an accidental involvement of the fibrous tissue in this situation It always is associated with periarticular or intramuscular fibrositis near the affected nerve and elsewhere It produces pain along all or part of the affected nerve It is particularly likely to involve the intercostal sciatic and brachial nerves

EXAMINATION There is no demonstrable synovitis or intra articular disease no juxta articular atrophy of muscles no loss of weight and no roentgenographic evidence of arthritis

On examination of the affected areas there will be found tender nodules cords or bands in the muscles fascia or articular capsule in about one half of these patients The concentration of hemoglobin remains at its previous level and the sedimentation rate remains normal or is only very slightly elevated

DIAGNOSIS Stiffness that appears in the morning but is relieved by moderate activity aggravation of symptoms by exposure and by changes of w

movements that do not occur normally in the joint In unimpacted fracture of the neck of the femur the lower extremity can be shortened by compression in the long axis or lengthened by traction in this axis Such abnormal mobility is associated with local pain absence of such pain points either to an old lesion or if recent to analgesia due to tabes or to syringomyelia

Palpation may be extended and refined by the use of a probe in case a fistula or a sinus exists On probing if the bone is bare the raw bone can be felt as grating if it is insensitive the bone is necrotic

Ossification of Bones A knowledge of the normal times for the completion of ossification is important in the diagnosis of many bone disorders of younger patients

Apart from the clavicle and the majority of the skull bones which are ossified in membrane the others undergo intracartilaginous ossification Interference with the ordinarily regular sequence of ossification of the bone may result from hereditary congenital or acquired causes The resulting diseases are all rare and certain ones affect membrane bones while others affect only those bones preformed in cartilage

Osteogenesis Imperfecta Osteogenesis imperfecta also designated fragilitas ossium brittle bones osteopsathyrosis and Lobstein s disease is a rare disease of bones manifested by the presence of multiple fractures after slight trauma Many instances are associated with blue scleras

Bickel has classified all forms of osteogenesis imperfecta somewhat as follows (A) *Hereditary Type* (1) Blue sclera and characteristic roentgenologic changes (2) white sclera and characteristic roentgenologic changes (3) blue sclera and atypical roentgenologic changes (B) *Nonhereditary Congenital Type* (1) Blue sclera and typical roentgenologic changes (2) white sclera and typical roentgenologic changes

It is impossible to distinguish clinically between the hereditary and the nonhereditary congenital types of osteogenesis imperfecta There is a great variation in the severity of the process from a disease in the newborn which is so severe as to be incompatible with life to a suspected condition which proves to be unrelated in a patient who is osteogenically healthy

These patients are poorly nourished short in stature and deformed The head tends to be triangular in shape with an increase in its transverse diameter The skulls of infants may feel like parchment the sutures are open Dentition is poor and the teeth often are soft and carious with a translucent appearance

The axial skeleton is most severely and most markedly affected Scoliosis and kyphosis are frequent and severe Deformity of the thoracic cage varies and the ribs and sternum often are twisted and molded to conform to the deviations of the spinal column

The deformity of the extremities is dependent on the number of fractures and on the success of alinement of the fragments of bones In some cases the magnitude of the problem of multiple fractures defeats all attempts at treatment

Excessive mobility of joints attributable to unusual laxness of the capsule and ligaments may be present These individuals are prone to have frequent sprained joints and tears of joint ligaments

Multiple fractures caused by trauma as slight as rolling over in bed are a constant feature The amount of pain suffered at the time is less than normal In Bickel s series there was an average of 9 2 fractures per case of the hereditary type and of 13 1 fractures per case of the nonhereditary congenital type

In cases of the *nonhereditary* congenital type the fractures occur earlier are more frequent and are caused by less trauma than in cases of the hereditary type of osteogenesis imperfecta The fractures are subperiosteal with little displacement of the ends of the bones but with angulation at the site of the fracture Many incomplete fractures or regions of absorption (Looser s zones) are revealed roentgeno

ing of the pelvis, scoliosis and limping A shortening of the radius or a lengthening of the ulna will cause radial flexion of the hand

When the bones are softened as in a severe degree of osteoporosis or when there has been solution of continuity from fracture the function of a bone as a supporting organ can no longer be fulfilled On attempting to use the bone as sup port so much pain is caused that the attempt is given up When pain is not felt by a child the child likely is congenitally insensitive to pain When pain is not felt by an adult syringomyelia or tabes dorsalis is suspected

Local enlargements connected with the skeleton are immobile a point which helps to distinguish them from masses near the bone, though an external mass may become adherent to an underlying bone and thus be rendered immobile for instance a carcinoma of the breast may become adherent to a rib

The form and the consistency of masses attached to bones or originating in them can often be felt on palpation In unimpacted fracture crepitation may be palpable When tumors or cysts originate within the bone and lead to atrophy of the bony substance over them this substance may be reduced in thickness to a thin paper like layer that crackles on pressure

In subperiosteal hemorrhage such as that seen after trauma and in infantile scurvy or sometimes in cephalhematoma new bone may form at the periphery On palpation a raised circular deformity of bone will be perceptible which as the pal pating hand passes toward the center of the mass suddenly ceases giving the im pression of a defect in the bone

On palpation over tumors of bone especially angiomas pulsation can often be felt On palpation of the skull if a pulsation is observed it may be due either to a bony tumor or to pulsation of the brain felt through a defect in the bone

In fractures of bones palpation by trained fingers will yield clues to the exact nature of the deformity and displacement In diseases of the upper cervical portion of the spinal column palpation through the pharynx gives access to the vertebral bodies In diseases of the pelvic bones palpation through the rectum or through the vagina may be helpful in diagnosis

When crepitus is perceived near a joint the examiner determines whether it is in the joint itself or due to a fracture of bone or to separation of the epiphysis near the joints In crepitation due to a fractured rib the crepitation may be audible during the respiratory movements Fractures are not always accompanied by crepitation since the ends of the bones may not be in apposition owing to marked separation or to erosion of the ends of the bones from tumor or again crepitus will be absent if the fracture is impacted

In testing the bones for tenderness gradually increasing pressure and tapping with the fingers may be used By such tests over long bones and on the spinous processes of the vertebrae disease of bones and of the spinal column is often localized

Tenderness of the bones may be diffuse as in leukemia in the state of exhaustion that follows influenza and rubella and especially in osteomyelitis or it may be strictly localized as in fractures or in bone abscesses Bone tenderness is commonly present in the neurotic patient

An important method of eliciting bone tenderness is by compression in the long axis, thus Press upward from the elbow toward the shoulder in testing the humerus from the heel toward the pelvis in testing the lower extremity or from the head or shoulders toward the pelvis in testing the spinal column In case of suspected im pacted fractures manipulation is carefully performed lest an impaction may be undone which might be desirable to keep

Mobility where normally there is none is a sign of solution of continuity and points either to unimpacted fracture or to pseudarthrosis To discover such abnor mal mobility in the neighborhood of joints the examiner should try to produce

INFECTIONS OF BONES

(Inflammatory Osteopathies)

The infections of the bones comprise osteitis osteomyelitis and periostitis

Osteitis Acute osteitis occurs in conjunction with various bone infections In the acute form osteitis is rarely a separate entity

Chronic osteitis is commonly diagnosed by the roentgenologist The clinician can rarely find supporting diagnostic evidence that chronic osteitis is a separate entity The term also is used to denote diverse bone conditions of noninfectious origin

As a result of infections there often occur (1) atrophy of disuse (2) subsequent deformities as the result of healing (3) epiphyseal defects separations and instability (slipping) and (4) exostosis During the process of a low grade infection a fracture may occur as during syphilitic infections of long bones

Osteomyelitis Osteomyelitis may be classified according to its manifestations as (1) acute (primary) (2) secondary and (3) chronic

Common sites are the shaft of the femur the clavicle scapula a rib and the tibia Any bone may be involved however including the bones of the skull the spinal column the sternum and the small bones of the extremities If the infection of the bone is a virulent one acute abscesses usually form in and around the affected bone Often a considerable mass of bone dies (necrosis of bone) and the sequestrum will keep up chronic suppuration with sinus formation unless an operation is performed

Acute Osteomyelitis Acute osteomyelitis is common in childhood but is rare in adult life It is a hematogenous infection of bone often due to the staphylococcus The infection may arise from some pre-existing lesion such as a furuncle septicemia or otitis media However more commonly it arises through some occult or unknown portal of entry There is frequently a history of injury at the site of the infection but its etiologic importance may not be established

Pus under the periosteum strips the membrane from the bone thus forming a subperiosteal abscess which ruptures into the overlying soft tissue and forms a sub cutaneous abscess The amount of bone infected by the disease is not immediately apparent In severe osteomyelitis when the infection involves the whole medullary cavity the entire shaft may become necrotic In other instances the infection may break through the cortex so soon that very little bone is invalidated Although the infection rarely penetrates the epiphyseal line it may precipitate a pyogenic arthritis by entering the joint from without after pus has appeared under the periosteum and extended from there into the joint

There are two reparative processes First occur the separation and partial phagocytosis of necrotic bone which becomes demarcated from the viable cortex and is called a sequestrum Second osteogenesis occurs from the lifted up periosteum although the new bone involucrum cannot be revealed roentgenologically for two or three weeks This new bone which develops in the presence of the sequestrum eventually generally becomes dense hard and rich in calcium and replaces the necrotic bone On rare oc casions the infection does not extend or penetrate but forms a localized lesion in bone and is called *Brodie s abscess* which ordinarily develops insidiously and is really a mani festation of chronic osteomyelitis From the primary bone lesion metastatic foci may develop and it is not uncommon for several bones to become infected one after the other This succession may continue for years so that eventually most of the long bones become invaded by the osteomyelitic process Abnormal lengthening of the affected bone occurs in many children after many months and is apparently due to the increased blood supply the hyperemia at the epiphyseal line augmenting local bone growth Owing to the preferences of the osteomyelitic process for the tibia femur humerus radius ulna fibula and metatarsal bones the irregularities in length do not cause grave locomotor difficulties Locomotor disturbances are consequences of the active disease

SYMPTOMS The general symptoms are those of general septicemia with fever chill rapid pulse and prostration The extreme pain seems to enhance the prostra

graphically The frequency of fractures ceases as puberty is approached and fractures may stop after puberty

From the foregoing discussion it is clear that only in the rare case without a history of multiple fractures from birth on is osteogenesis imperfecta likely to be confused with osteitis fibrosa generalisata The bone disturbance may be first recognized at the age of about 15 or 16 years when roentgenograms reveal a generalized lack of bone density The very thin but not decalcified bone rules out the bone disease of hyperparathyroidism The lack of bone density may or may not be accompanied by blue scleras *The concentrations of serum calcium and serum phosphorus* are within normal limits The concentration of serum phosphatase may be slightly elevated There probably will be a family history of fractures blue scleras or otosclerosis Blue scleras and otosclerosis are not constant features of this *disease*

Osteochondrosis of the Capitular Epiphysis (Legg Perthes Calve Disease Osgood Schlatter Disease Kohler s Disease) Osteochondrosis of the capitular epiphysis (Legg Perthes Calve disease coxa plana) is a disease of the upper part of the femur affecting children between 5 and 10 years of age This disease has been thought to have an infectious element in its etiology and thus the designation osteochondritis deformans juvenilis has been used

Symptoms comprise slight pain and limp that are soon followed by shortening of the leg This disease may symptomatically resemble coxa vara

Diagnosis is established by roentgenographic examination The roentgenograms reveal progressive flattening of the head of the femur which is characteristic of the disease A similar lesion has been described in the tibial tubercle (Osgood Schlatter disease) and in the tarsal scaphoid (Kohler s disease) Treatment consists of immobilization when pain is present

Lipochondrodystrophy (Gargoylism or Hurler s Syndrome) This is a rare condition ending in death in childhood Mental deficiency is associated with malformations of the bones There is dwarfish stature with short neck curved spinal column short thick limbs, protuberant abdomen and grotesque face The nature of the facial peculiarities somewhat resembling those of cretinism is not clear Optic atrophy may occur In many cases enlargement of the liver and the spleen gives rise to suspicion of congenital syphilis The corneas may exhibit opacities in the deeper layer

Periodic Arthralgia of Hereditary Origin Periodic arthralgia is characterized by regular recurrences of pain and often *swelling of joints of the extremities* but usually of one or both knees It is a rare disease The disease has been known to affect five generations of the same family (Reimann)

The disorder has the characteristics of the Schonlein Henoch syndrome Both probably are manifestations of the same underlying cause and represent variant forms of a group of periodic diseases The disorder may begin at any time of life and the intervals may shorten or lengthen It may last a lifetime or may disappear either spontaneously or in rare instances after certain forms of therapy Rarely do permanent injury to the joints and other effects on the general health occur The episodes usually recur at regular intervals of 7 to 21 days last several days usually without fever or other symptoms or signs, and disappear until the next predictable bout In some patients recurrences come at irregularly short or long intervals In some instances there is only stiffness and in others there is an effusion into the joint or joints

Biopsy reveals edema of the synovial membrane with or without monocytic cell infiltration of the tissue Effusion of serous fluid when present may contain more than 1 000 cells per cubic millimeter either polymorphonuclear cells or lymphocytes predominate

Osteomyelitis of the skull is secondary to compound fracture or a sinusitis Osteomyelitis is always suspected when an abscess or infected wound located near bone fails to heal and forms a chronic draining sinus

Roentgenologic examination is required in order to make a diagnosis in secondary osteomyelitis However the diagnosis is usually evident from the history the findings and the situation and appearance of the lesion

Chronic Osteomyelitis Chronic osteomyelitis follows an acute primary or secondary osteomyelitis which has resulted in necrosis of bone The severity of chronic osteomyelitis often depends on the extent of bone killed by the virulence of the infection An extremity which is the site of chronic osteomyelitis shows considerable muscular and bone atrophy and exhibits one or more draining sinuses leading to the bone The patient is often shrunken from the toxemia pain and fever This disease often is caused by *Salmonella typhosa* after an attack of typhoid fever

Roentgenologic examination will reveal the extent of the lesion and often will clearly demonstrate the site and size of sequestra

Tuberculosis The distinction between tuberculosis of the bone and of the joint cannot always be made since the tubercle bacillus often attacks both structures simultaneously or seemingly so

Tuberculosis of the Cranial Bones The infection usually gains access to the middle ear through the eustachian tube and is secondary to infection of the throat cervical lymph nodes or lungs It may spread from more remote foci however by way of the blood stream

In most cases the process is a sluggish one but in others it progresses rapidly invading the mastoid bone and the labyrinth External pachymeningitis with penetration of the dura resulting in meningitis or formation of tuberculoma is common The sigmoid sinus may be thrombosed In a few cases tuberculous osteomyelitis of the base of the skull in the anterior fossa has been observed This is apparently due to extension of tuberculous processes in the sinuses and nasal bones

Tuberculous otitis is usually secondary to lesions in the upper respiratory passages The onset is gradual in most cases and is not accompanied by pain There is a thin watery discharge at first but this may later become purulent if secondary infection develops There may be multiple perforations of the drum and later the whole drum may be destroyed In some cases hearing may be only slightly reduced in many it is completely abolished A characteristic feature is the presence of paralysis of the seventh nerve which has been estimated to occur in 40 per cent of all cases Ford observed cases also in which osteitis of the walls of the orbit resulted in palsies of the third nerve

The infection may extend through the dura causing tuberculous meningitis or solitary tubercle The latter is most likely to be found in the cerebellar hemisphere or the temporal lobe In instances complicated by pyogenic infections abscess of the brain or purulent meningitis may result

The diagnosis depends on the evidences of tuberculosis elsewhere in the body the slow course absence of pain the multiple perforations of the ear drum the frequent presence of facial paralysis and above all on the demonstration of tubercle bacilli in the secretions and granulations

Tuberculous Spondylitis Tuberculosis of the vertebrae (Pott s disease) is the most serious inflammatory disease of the spinal column especially in children

The process is believed to begin in the body of the vertebra and extend to the periosteum the intervertebral disks and ligaments From 1 to 5 adjacent vertebrae may be affected and in some there are several foci in the spinal column As a result of weight bearing the bodies of the vertebrae may in some collapse causing an angular kyphos The spinal cord or the spinal nerve roots may be affected by an abscess of the vertebral body extending into the epidural space and compressing them Generalized or localized tuberculous meningitis may be present Compression of the cord may occur as a result of dislocation and deformity of vertebrae or of the dorsal displacement of a sequestrum

tion During the first day or two of the disease before the infection has broken through the cortex of the bone the pain is severe and it is situated over the end of the bone near the joint In adults the general reaction is likely to be less marked than in children subacute and chronic forms are more frequently encountered in which the infection develops slowly

Streptococcal osteomyelitis differs considerably in its manifestations from osteomyelitis produced by the staphylococcus The former infection much more commonly occurs as a metastatic invasion of the bone from a primary source such as otitis media or mastoiditis or from a furuncle or carbuncle The manifestations of the streptococcal osteomyelitis are likely to develop more insidiously than those of the staphylococcal infection The pain is not so severe swelling and subcutaneous suppuration develop more slowly and less commonly There is less destruction of bone and sequestration is therefore less marked Many such lesions will subside under proper conservative care without gross suppuration They are particularly prone to occur in infants

Examination Muscle spasm is often present The joint can be moved somewhat without increasing the pain Acute tenderness over the involved bone on deep palpation is one of the earliest signs In eliciting this tenderness the bone is carefully palpated starting at some distance from the lesion which is approached gently and gradually The overlying tissues may be swollen and edematous but the skin itself is not red or tender during the early stage of the disease If the infection has been present for several days there may be evidence of a local abscess that is swelling redness local heat and fluctuation

Diagnosis The diagnosis is based on the foregoing findings The local signs and symptoms are meticulously observed Examinations are repeated at short intervals in order to keep informed in regard to the rapidly developing events The results of roentgenologic examination are disappointing because the earliest roentgenologic evidence of acute osteomyelitis is absorption of bone which ordinarily is not demonstrable for a week or more after the onset The diseases which may be diagnostically confusing are acute rheumatic fever acute pyogenic arthritis and acute cellulitis Involucrum formation requires at least 2 weeks It is discovered on roentgenologic examination when calcium has become deposited under the elevated periosteum Sequestration may not be demonstrable for an even longer period

When acute osteomyelitis is suspected a blood culture which will often reveal the bacterial cause should be made It is important to remember that in osteomyelitis during adolescence the urine may contain the etiologic agent

In the differential diagnosis acute osteomyelitis is distinguished from (1) acute arthritis especially from acute coxitis (2) subcutaneous abscess and intramuscular abscess (3) acute rheumatic fever (4) ordinary septicemia and pyemia and (5) malarial fever

In general operation is urgent in staphylococcal infections not urgent and frequently unnecessary in streptococcal infections

Secondary Osteomyelitis This is a bone infection occurring as the result of infection in tissues adjacent to the bone for instance such as that due to compound fractures

Secondary osteomyelitis of the jaw often develops by extension from cellulitis about a tooth If the tooth is pulled at the improper time for instance while the infection is too acute osteomyelitis is particularly prone to develop and may lead to septicemia

The terminal phalanx may become the site of osteomyelitis during the course of *a felon* The other phalanges may be infected secondarily from acute suppurative tenosynovitis Osteomyelitis of the tarsus is sometimes secondary to penetrating infections and may eventually involve the entire foot The patella and olecranon are sometimes involved by direct extension from an overlying lesion

Granuloma Inguinale Granuloma inguinale of bones occurs during the course of the active infection The skeletal infection is characterized roentgenologically by osteolytic areas in the bones In none of the cases so far reported are the roentgeno logic changes specific for this disease Diagnosis is best established by biopsy with search for macrophages containing Donovan bodies (*Klebsiella granulomatis*) when the bone changes occur during the course of granuloma inguinale (see Diseases of the External Female Genitalia Chapter 7)

Syphilis Syphilis of the bone may be manifested as chronic periostitis Syphilitic osteitis and periostitis or combined syphilitic bone lesions may be present either in hereditary syphilis or in acquired syphilis In the latter the bones may be involved either in the secondary or in the tertiary stage of the disease

During the secondary stage of syphilis a patient who has syphilis of the bone may complain of rheumatic or of neuralgic pains or of circumscribed tenderness on the bones The superficial bones (skull anterior surface of tibia ulna metacarpi) are most often painful The pains are milder in the daytime than at night when they may become almost unbearable Nodes due to subperiosteal deposits often be come palpable

The usual affection of the bones by syphilis is periosteal gumma formation with necrosis of the underlying bone followed by ulceration and exposure through the skin or by an extensive osteophyte formation Less common is gummatous osteitis or osteomyelitis with necrosis and erosion of bone

Syphilitic inflammation of the bone may produce a new growth of spongy bone on the surface of the cortex Such proliferations of bone may form a convex layer on the bone cortex which as time passes becomes very hard and solid However these cortical layers of proliferated bone are not limited to syphilitic infections for other nonspecific forms of periostitis can produce the same thing All of these pro liferating bone disturbances occur more commonly in the skull and long bones especially the tibia than in other bones

Gummatous osteitis and osteomyelitis may arise in the interior of the bone In the cranial bones the periosteum can be separated by such a process leaving a coarsely corroded surface If the process is extensive the skull may be penetrated and destroyed and left with a ragged hole in it Comparable gummatous nodes may occur in the tibia extending into the cortex and marrow cavity which may circum scribe the bone and cause necrosis of it Gummatous lesions may occur within the marrow cavity and involve it locally or may extend to involve the whole of it without outward evidence of their presence In other instances marrow involvement will be accompanied by a spindle shaped enlargement of the shaft of the bone

In syphilitic dactylitis there is osteoperiostitis with enlargement and rarefaction and internal destruction of the bone The finger swells and may be easily fractured and subsequent fistula formation ensues When a phalanx or a metacarpal bone is affected the appearance may resemble that of tuberculosis

In roentgenograms the signs of syphilis of the bone are usually characteristic Sometimes a circular involvement of the bone is recognizable A positive serologic reaction may not be helpful diagnostically a negative serologic reaction is definite in that the diagnosis of syphilis of the bone should not be made if the reaction is negative

Syphilis of the bones may resemble tuberculosis sarcoma sporotrichosis or even chronic staphylococcic osteomyelitis

Yaws Yaws is caused by *Treponema pertenue* In this disease periostitis affects many bones causing underlying necrosis of bone and sclerosis

The symptoms cannot be distinguished from those of syphilis Multiple tendon sheaths are involved which give joint symptoms but the joint surface is not affected so that primary joint symptoms are not present

The serologic reactions are positive The roentgenologic findings are very much

The spinal nerve roots may be involved by infiltration or by compression when there is collapse of the vertebrae and shortening of the spinal column

SYMPTOMS The onset is insidious and is characterized by pains in the distribution of the spinal nerve roots arising in the affected region of the spinal column In children sleep is broken by night cries as the result of relaxation of the muscle spasm of the vertebral muscles

The pain is usually referred around to the anterior surface of the body Among other symptoms characteristic of the early stages of the disease are abnormalities of posture which result from fixation of the spinal column in positions adapted to relieve the pressure on the affected vertebrae Such fixation affects posture station and gait The patient walks carefully with a rigid back fearful of jarring it There are loss of weight fatigue and perhaps afternoon rise of temperature

EXAMINATION When the cervical vertebrae are involved the pain is referred to the occiput neck arms or shoulders depending on situation of the lesion The head may be held rigid flexed extended tilted or rotated to one side There is strong muscle spasm and rotation of the neck is resisted If abscess forms it may drain into the retropharyngeal space or less commonly laterally into the muscles of the neck

Lesions of the thoracic vertebrae cause the body to be inclined forward The lumbar lordosis is diminished The patient walks with caution often on the toes to avoid jars When the upper thoracic vertebrae are affected the shoulders are often elevated and squared If abscess forms it drains into the posterior mediastinum and causes dyspnea and cyanosis

Lesions in the lumbar vertebrae cause increased lordosis Psoas abscesses may cause psoas spasm with flexion of the hip

Ford has emphasized that paralysis due to compression of the cord by epidural tuberculous tissue may be present without deformity of the spinal column without muscle spasm and without demonstrable changes in the roentgenograms of the bones

Involvement of the spinal cord is from pressure manifested only by weakness and stiffness of the legs without paraplegia or loss of control of the bladder In instances of lesions below the lumbosacral enlargement paralysis if present is associated with atrophy flaccidity and loss of reflexes

DIAGNOSIS The diagnosis is based on the presence of loss of weight retarded growth fever changes in gait altered posture and night cries muscle spasm and limitation of movement deformity of the spinal column positive intracutaneous tuberculin reaction and the roentgenographic changes in the spinal column In an occasional case there is paraplegia However 8 of every 10 who have this sort of paraplegia recover in less than one year In some recovery may be expected after several years

Periostitis Periostitis is an inflammation of the covering of the bone which occurs in both acute and chronic forms Acute inflammation of the periosteum like osteitis occurs in association with various disease conditions of bone and as such cannot be definitely diagnosed except by means of roentgenologic examination which reveals the characteristic increased density about the cortex of the bone

Acute Periostitis Acute periostitis follows either trauma as in fracture or contusion or infection as in acute periostitis of the jaw secondary to dental infection An acute periostitis is not uncommon as a complication of severe fevers

There are local swelling extreme pain and heat When sepsis is present suppuration may follow When the periostitis is of purely traumatic origin the condition is usually transitory

The diagnosis is usually easy if the etiology is known Roentgenologic examinations will differentiate acute from chronic periostitis since in the latter there will be evidence of bone formation

active person in plaster casts an acute osteoporosis may arise An acute osteoporosis is more likely to arise if the patient ingests large amounts of milk and if in order to accelerate healing of the bone vitamin D is administered When the foregoing con ditions prevail the excessive amounts of calcium cause a hypercalcemia and hypercalciuria The hypercalciuria is the cause of the formation of *renal stones* which occasionally occurs in patients who are in extensive casts Renal stones seem to be of a higher incidence in those who have fractures of the pelvis and the femur The formation of renal stones is less frequent in patients who have fractures of the upper extremity

Anuria in a patient who has extensive immobilization of the skeleton occasion ally occurs If anuria does occur the urine and the blood are examined for an excess of calcium If there should be an excess of calcium the removal of the cast may be a lifesaving measure

In such patients the high calcium content of the serum and the urine may give the impression of hyperparathyroidism Hyperparathyroidism is a rare disease and its differentiation need not delay the removal of the cast

Osteoporosis and the Senium (*Senile Osteoporosis*) A characteristic of senility is decreased intensity of protein synthesis manifested by atrophy of skin and muscles and by osteoporosis As the years advance beyond middle age often the cortex of the bones of the whole skeleton shows signs of atrophy The trabeculae of the cancellous bone become thinner and further apart the greater part of the bone marrow becomes fatty and only a few scattered islands of red bone marrow are in evidence In extreme examples of the disease the vertebrae collapse and the cortex of the other bones may become thin This is the commonest form of osteo porosis The skull and extremities are practically always free from osteoporotic involvement except in the severest forms of the disease and the lamina dura of the teeth remains normal The vertebrae are the most profoundly affected

Osteoporosis and Vitamin C Deficiency In children the commonest evidences of the lesions of scurvy are the irregular broadened epiphyseal line the lateral spurs and occasional dislocation of the entire epiphysis the decreased calcification behind the epiphyseal line the thin diaphyseal cortex and the rarefied shaft Normal calci- fication of bones and teeth seems to be dependent on adequate vitamin C Ascorbic acid deficiency leads to decreased bone matrix formation and in turn to *osteoporosis*

Adult Scurvy Scurvy is rare in adults The bone lesions in the adult are limited to subperiosteal hemorrhages as described in infantile scurvy Intracranial bleeding is spoken of but it is not common in the adult

The history of deprivation of those foods which contain vitamin C is usually definite except when scurvy occurs during the course of chronic starvation The concentration of vitamin C is decreased in the blood Roentgenologic examination of the long bones may be diagnostic

Osteoporosis and Rickets Rickets is a metabolic disease due to deficiency of vitamin D caused by insufficient exposure to sunlight which produces changes in the viscera muscles and tendons The characteristic changes in the disease referable to the skeleton which take place at the periods when the skeleton is developing most rapidly (that is during the first two years of life) extend through to the age of puberty

When there is insufficient absorption of calcium from the intestine proteinic bone matrix is formed in the normal way but no calcium phosphate is available for deposition As the normal growth or wear and tear of the skeleton go on there is not enough calcium for growth or the lost calcium is not replaced and decalcifica tion or osteoporosis appears (see Chapter 22)

Osteoporosis and Osteomalacia The reactions of the skeleton in rickets and osteomalacia are fundamentally identical In both diseases the osseous trabeculae are surrounded by broad osteoid seams, and thus the definition of osteomalacia as

the same as those of syphilis The bone lesions are benefited by specific treatment for syphilis The diagnosis depend more on the age of the patient who as a rule is much younger than one who has syphilitic bone disease It is well not to make a diagnosis of yaws of the bone unless the patient comes from an area in which the disease is *endemic*

Hydatid Disease Hydatid disease of bone occurs in only a few of those who have hydatid infection The common sites affected are the vertebrae the pelvis the femur and the humerus The vertebrae are the bones most frequently affected The *spongy tissue of the vertebral bodies* is a favorite site when hydatid disease attacks a bone The infection as with hydatid disease elsewhere usually starts in childhood While it is confined to the bone the growth of the hydatid is so slow that symptoms do not become evident before adult life There are no symptoms until the destruction of the bone leads to fracture or dislocation or until the hydatid cyst has erupted from the bone and caused *pressure or swelling*

The diagnosis of vertebral hydatid disease is uncertain The general condition of the patient is usually excellent Symptoms are due to fracture or subluxation of the affected vertebra or to pressure by extraosseous extensions which may be confused with lipoma or cold abscess Although vertebral hydatid disease has no pathognomonic roentgenologic signs certain features such as absence of periosteal reaction minimal collapse slight involvement of intervertebral spaces only increased density of bone together with areas of destruction and involvement of contiguous ribs when a thoracic vertebra is affected aid the roentgenologist in the interpretation of the roentgenogram

METABOLIC DISEASES OF THE BONES

Metabolic diseases of the bones may arise during the course of malnutrition or as the result of deficiencies of vitamins

Albright has reasoned that the diet is of etiologic importance in the production and the maintenance of bone If there should be inadequate amounts of protein ingested the osteoblasts would be deprived of the necessary strength to produce the organic matrix which is the first requisite for the formation of bone To the contrary if there is an increased availability of calcium and phosphorus in the system as a result of a diet rich in these substances there would be a decrease in normal degree of resorption of bone However reasonable these assumptions are there is no evidence that osteoporosis ever results from an inadequate diet in man

Osteoporosis and Metabolic Disorders of the Bones Osteoporosis is a disease of the bones characterized by increased porosity from widening of the haversian canals and absorption or lack of calcareous material Osteoporosis occurs in many and diverse conditions which permit a decreased formation of proteinic bone matrix Osteoporosis can be caused by any condition in which the synthesis of protein is impaired In such instances the calcium taken from the intestine cannot be used for normal bone repair because no bone matrix has been formed It therefore is excreted in the urine Thus in the initial stages of osteoporosis hypercalciuria is a frequent occurrence and occasionally renal stones develop Osteoporosis occurs frequently in prolonged *immobilization of bone* in *metabolic disturbances* and *deficiency states* in the *senium* in *prolonged semistarvation* in the *menopause* and in *Cushing's syndrome*

Immobilization of Bones The daily use of the skeleton requires a certain amount of maintenance The maintenance of the skeleton is a function of the osteoblasts During immobilization of the skeleton the normal stimuli for osteoblastic activity are not operative less proteinic matrix is made by the osteoblasts and thus less bone is formed In time often in a week or two the limb in a plaster cast reveals atrophy of the soft parts and particularly atrophy of the bone

In instances of immobilization of a large part of the skeleton of a previously

Polyostotic Fibrous Dysplasia Polyostotic fibrous dysplasia in females is characterized by abnormalities of skin and bone and of endocrine function The cutaneous lesions are brown nonelevated pigmented areas which tend to be on the same side as the osseous lesions The osseous lesions on histologic examination reveal osteitis fibrosa

SYMPTOMS The first manifestation of the disease is a segmental distribution roughly similar to that of the osseous lesions of pigmented areas of skin which consist of small or large irregular cafe au lait spots These lesions are often first observed soon after birth and are similar in every respect to the forme fruste of Recklinghausen s disease

The skeletal manifestations are varied They may be without symptoms their presence having been discovered accidentally on roentgenologic examination on account of a fracture The fracture is therefore not a true one but a pathologic fracture The symptoms are often grouped according to the grouping of the osseous lesions that is there may be pains in one leg and foot one arm and hand and headache

The symptoms of hyperthyroidism may develop at an early age

Female patients who have osteitis fibrosa disseminata often exhibit sexual precocity Catamenia may be established irregularly at an early age in some instances before the age of 5 years The breasts develop and pubic and axillary hair appears Synchronous with the sexual precocity is an associated skeletal and somatic precocity During childhood these patients grow rapidly and are large for their chronologic ages but growth ends before a large size is attained owing to early closure of the epiphyses

EXAMINATION These children are large for their age If adulthood has been obtained the patient is short of stature The pigmented areas are present over an extremity in which lesions and fractures of bones are present The forehead is often prominent As stated in female patients the breasts are well developed and pubic hair and axillary hair are present during childhood The thyroid gland often is easily palpable

Involvement of the upper ends of the femora often results in outward bowing which in the presence of the bone disease predisposes to recurring fractures and pseudarthroses

Roentgenologic examination reveals osseous lesions characteristic of polyostotic fibrous dysplasia These lesions are usually confined to one side of the body except in the skull where there may be a bilateral thickening of the bones at the base with obliteration of the frontal and sphenoidal sinuses The bone age is greater than the normal or actual age of the patient

The basal metabolic rate is often increased Serum calcium serum phosphorus and serum alkaline phosphatase are usually within normal limits of concentration In these precocious girls the values for 17 ketosteroid urinary excretion and follicle stimulating hormone are within the range of values for adult menstruating women In boys or men who have the disease hormone disturbances have not been described

DIAGNOSIS The history the physical examination and the presence of extensive osseous lesions which show an increased density on roentgenologic examination are sufficient for a diagnosis

Postmenopausal Osteoporosis Between the ages of 40 and 60 years osteoporosis occurs almost exclusively in women It has therefore been assumed that the osteoporosis which develops in the 40 to 60 year age group may well be closely related to postmenopausal (endocrine) processes

The assumption that osteoporosis in women more than 45 years of age is related to the postmenopausal syndrome is based on the proved influence of estrogenic hormones

adult rickets is justified In both conditions the osteoporosis as revealed by roentgenologic examination is definite and sometimes *of an extensive degree*

ENDOCRINE DISTURBANCES OF BONE

Disorders of the endocrine glands may profoundly affect the formation and the maintenance of bones Of these hormonal disorders the work of Albright has revealed that hyperparathyroidism is the most spectacular example

Osteitis Fibrosa Cystica of Hyperparathyroidism This bone disease is characterized by severe bone pains generalized osteitis fibrosa skeletal softening with spontaneous fractures angular deformities of the bones and cysts and cystic tumors of the bones The tumors are often referred to as giant cell tumors There are an increased concentration of *calcium in the presence of decreased concentrations* of inorganic phosphorus and an increased concentration of alkaline phosphatase in the serum There is an increased urinary content of calcium and phosphorus In these patients bilateral renal stones and disturbed renal function may be present The disease occasionally is accompanied by muscular hypotonia fatigue polyuria palpitations and dyspnea Nausea and vomiting are common The bone changes are easily demonstrated by roentgenographic examination

On examination tumors may be found in the jaw metacarpals, metatarsals and at the ends of long bones Bone deformities include bending of the long bones deformities of the pelvis similar to those seen in osteomalacia and various deformities of the vertebrae A result of the vertebral changes is a decrease of stature a pigeon breast deformity of the thorax and a disappearance of the neck into the thorax The bones in this disease are more brittle than in osteomalacia and fracture rather than bend The patient may be first examined for a recent fracture

Diagnosis of osteitis fibrosa cystica can be established on roentgenologic examination by the presence of a generalized decalcification and the even ground glass appearance of the skull cysts and tumors A cortical cyst is most suggestive of hyperparathyroidism The teeth fail to show any decalcification and therefore contrast markedly with the decalcified bones of the face The diagnosis is confirmed by the presence of hypercalcemia hypophosphatemia and excessive urinary excretion of calcium

Secondary Hyperplasia of the Parathyroid Glands and Bone Disorders In this condition the parathyroid glands respond to the abnormal calcium and phosphorus metabolism rather than initiate it

Secondary hyperplasia of the parathyroid glands most commonly occurs in osteomalacia and in multiple myeloma and occasionally in generalized malignant metastasis in the bones Secondary hyperplasia of the glands may occur in children and in adults who have chronic renal failure with acidosis The renal lesion seems to be one of the lower nephron type The disease is much more frequently present in those who have congenital lesions of the kidneys and changes in the skeleton

The bone lesions present under these circumstances consist of osteitis fibrosa and are designated as *renal osteodystrophy* Occasionally a certain amount of bone deposit is present around the bone trabeculae in those who have chronic uremia Despite the occasional occurrence of osteoid zones the designation of renal rickets is not considered desirable by Snapper He objects to the term renal rickets even in children and young adolescents who have chronic renal failure when the closure of epiphyseal disks is delayed and roentgenographic changes resembling those of rickets are seen Snapper prefers the term renal osteofibrosis or osteonephropathy

The manifestations of *secondary hyperparathyroidism* in chronic renal failure are hypocalcemia acidosis and usually hyperphosphatemia In instances of extensive parathyroid hyperplasia the calcium of the serum is often normal or low normal It seems possible that the secondary hyperplasia of the parathyroid glands is responsible for the change from the original hypocalcemia to a normal calce

The thickening of the cortical and spongy tissue in some typical places such as the skull ribs vertebrae and femora may be so pronounced that macroscopic inspection discloses no difference between the cortex and the spongiosa The long bones are characteristically clubbed in shape

The symptoms are variable There is a retardation in physical development but mental symptoms may or may not be present Often optic atrophy and blindness are the first symptoms Repeated bone fractures are characteristic The head may be enlarged probably owing to the thickening of the cranial bones as well as to a slight hydrocephalus

On roentgenologic examination bands running parallel to the line of the epiphyses may be observed in the bones of the hands and feet and in the metaphyses when there are only moderate degrees of calcification In the scapulae the ossa ilii and the tarsal and carpal bones these bands are ring-shaped

Biochemical tests reveal no signs of hyperparathyroidism The blood has normal values for calcium inorganic phosphorus and magnesium There is a normal number of units for (alkaline) phosphatase The cellular elements of the blood are normal in the early stages of the disease The absence of anemia has been regarded as an argument in favor of the conception that the osseous anomalies of marble bone disease are primary and not dependent on the damage of the bone marrow

In favor of the diagnosis of marble bones is the presence of atrophy of the optic nerve associated with the tendency to bone fractures malformation of the thorax the thickening of the lower end of the femur the peculiar shape of the head and deformed or incomplete teeth

Chalky Gout (Tophaceous Gout) Chalky gout is a form of calcinosis according to Rosenberg characterized by deposition of calcium salts in the terminal phalanges of the toes The mechanism by means of which the calcium is deposited is unknown The calcium concretions consist of calcium phosphate and calcium carbonate which usually are present in the same proportion as in bone Small amounts of magnesium carbonate and phosphate cholesterol and fatty acids also have been found in the deposits

For a discussion of calcinosis see Calcium in Chapter 22

Osteitis Deformans (Paget s Disease) Sir James Paget wrote in part of osteitis deformans

It begins in middle age or later is very slow in progress may continue for many years without influence on the general health and may give no other troubles than those which are due to the changes of shape size and direction of the diseased bones

'The disease affects most frequently the long bones of the lower extremities and the skull and is usually symmetrical The spine by change in its own structures may sink *and seem to shorten with greatly increased dorsal and lumbar curves the pelvis* may become wide the necks of the femora may become nearly horizontal but the limbs however misshapen remain strong and fit to support the trunk

In its earlier periods and sometimes through all its course the disease is attended with pains in the affected bones pains widely various in severity and variously described as rheumatic gouty or neurologic not especially nocturnal or periodical It is not attended with fever No characteristic conditions of urine or faeces have been found in it It is not associated with syphilis or any other known constitutional disease

'The bones examined after death show the consequences of an inflammation affecting in the skull the whole thickness in the long bones chiefly the compact structure of their walls and not only the walls of their shafts but in a very characteristic manner those of their articular surfaces

'The outer surface of the walls of the bones was irregularly and finely nodular as with external deposits or outgrowths of bone deeply grooved with channels for the larger periosteal blood vessels finely but visibly perforated in every part for transmission of the enlarged small vessels Everything seemed to indicate a greatly increased quantity of blood in the vessels of the bone

on calcium metabolism The decreased production of estrogens during the menopause may result in decreased osteoblastic stimulation

The symptoms of postmenopausal osteoporosis develop slowly However in some skeletal deformities with a marked loss of stature may be present The first symptom is often a sharp pain in the back after slight trauma On roentgenographic examination of the spinal column a marked decalcification, particularly of thoracic and lumbar portions is apparent Atrophy of the bone is often so advanced that only the end plates of the vertebrae are denser than the intervertebral disks In the lumbar region where the pressure is greatest in the center of the vertebrae the osteoporosis causes the formation of fish or hourglass vertebrae The elastic intervertebral disks expand at the expense of the softened vertebrae and often become wider than the vertebrae themselves Ultimately a complete collapse of one or more vertebrae may occur In less advanced instances there may be present herniation of the nucleus pulposus

Patients who have postmenopausal osteoporosis often show remarkable improvement after administration of testosterone propionate However actual recalcification of the osteoporotic bones rarely is observed

Osteoporosis in Cushing s Syndrome Cushing s syndrome may result from cancer or adenoma of the adrenal cortex unilateral or bilateral hyperplasia of the adrenal cortex or basophilic adenoma of the pituitary gland

In Cushing s syndrome the error in protein metabolism results in an inadequate repair of tissue manifested by atrophy of the muscular system and an insufficient formation of bone matrix and consequently osteoporosis The osteoporosis involves the spinal column pelvis and ribs The intervertebral disks compress the bony part of the spinal column and fish or hourglass vertebrae develop in the lumbar portion while wedging of vertebrae is prominent in the thoracic portion In the later stages of the disease actual compression fractures of the vertebrae occur In more advanced disease the skull and the long bones become osteoporotic In the calvarium the decalcification is localized mainly in the frontal and parietal bones

In the differential diagnosis hyperparathyroidism with osteitis fibrosa cystica generalisata lipoid granulomatosis (eosinophilic granuloma, Hand Schuller Christian disease xanthomatosis) and Recklinghausen s neurofibromatosis are considered

In those who have extensive involvement of the skeleton secondary lesions may arise which are incompatible with prolonged life The thorax may be so deformed that cor pulmonale is inevitable Pathologic fractures are common and usually heal well with the exception of those at the upper ends of the femora The endocrine disorder may be benign The skeletal lesions are usually benign Often there is little or no progression for a long time in the extent of the skeletal involvement

Hypothyroidism A characteristic of juvenile hypothyroidism is a retardation in osseous development with a resulting subnormal bone age as determined by roentgenologic examination In addition to the retarded development there may be some abnormal ossification of the cartilages of the epiphyses and of the round bones

DISEASES OF THE BONES ASSOCIATED WITH BLOOD DYSCRASIAS

Changes in the femora tibias fibulas and cranium of almost specific configurations occur during the course of some of the blood diseases such as Cooley s anemia Osteoporosis too may occur during the course of a chronic anemia (see Chapter 12)

DISEASES OF THE BONES OF UNKNOWN ETIOLOGY

Marble Bones (Osteopetrosis) The disease sometimes called also *ivory bones* and Albers Schonberg disease is often of familial occurrence (about 40 per cent of the cases) It may be hereditary

Complicating fractures may result from trifling causes

PSEUDOFRACTURES

Pseudofractures are transverse zones of rarefaction varying in width from less than 1 mm to more than 1 cm over which there is a thickening of the periosteum Pseudofractures affect various portions of the skeletal system and often are symmetrically distributed They are generally an indication of certain weaknesses or dysfunction of the skeletal system

There are no signs or symptoms which may be said to be characteristic of pseudofractures Pain is the commonest complaint and may be an ache or may be most noticeable when pressure is exerted over an affected region This is accompanied by a degree of limitation of function due in part to the pain elicited and in part to the subjective sensation of giving way of the affected bone When the leg and the pelvis are involved there may be a waddling gait similar to that found in congenital dislocation of the hip and coxa vara The patient may have the greatest difficulty in climbing stairs or in rising from the sitting position and may show a definite hesitation in beginning to walk The reflexes may be abnormal as a result of the disturbances in prolonged dysfunction of the calcium phosphorus metabolism

The diagnosis is made roentgenologically

TUMORS OF THE BONES

Bone tumors are classified as benign or malignant neoplasias

Benign Tumors Meyerding classified benign bone tumors as follows (1) osteoid osteoma (2) benign osteogenic tumors which occur as (a) osteoma or exostosis and (b) chondroma (3) benign fibroma (4) benign giant cell tumors (5) benign vascular tumors of bone (angioma) Also included in this category are the lymphangiomas and the fibrosis secondary to these processes (6) bone cysts and (7) epiphyseal chondromatous giant cell tumor (Codman s tumor)

1 **Osteoid Osteoma** An osteoid osteoma is a small benign neoplasm of bone consisting of osteoid tissues carried by a substratum of vascularized osteogenic connective tissue It occurs most frequently in boys and young men but it may occur throughout middle age The tumor is commonest in the tibia or femur but may occur in other parts of the skeleton The etiology is unknown

The first symptom is pain In the beginning mild and vague it progresses in severity and constancy and may be described as a boring localized pain It is worse at night than during the day If the lesion is near a joint there may be limping from the stiffness and weakness and pain on even limited motion Often when the patient first consults a physician the symptoms have been present for less than 6 months sometimes from 6 months to 2 years

There is a localized tender point and often a tumor in the painful area Disuse atrophy of adjacent muscles and increase in joint fluid simulating a primary arthritis may be present

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is established by histologic examination of the tissue obtained by biopsy or that removed at the time of operation

Osteoid osteoma is to be distinguished from solitary abscess sterile abscess of Brodie sclerosing nonsuppurative osteomyelitis and syphilitic or tuberculous osteitis

2 **Benign Osteogenic Tumors** (a) **Osteochondroma** (*Osteoma Exostosis*) An osteochondroma is a bony skeletal outgrowth with a cartilaginous cap The tumor occurs most frequently in boys and in men less than 25 years of age The tumors arise near the ends of long bones at sites of tendinous attachments The growth is outward in the direction of muscle These are usually congenital tumors

ETIOLOGY The etiology as stated by Paget is unknown The disease is one of advanced life setting in at any time after the age of 30 years

PATHOLOGY There is little to be added to the description of the pathology as this was originally made The gross changes consist of a general thickening and bowing of the long bones both characteristics being in evidence throughout the shaft and in advanced stages involving the epiphyses as well Not infrequently the bone is increased to twice its normal size In all cases the bowing is in the nature of an accentuation of the normal curve together with a moderate degree of torsion Actual lengthening also takes place The normal ridges and prominences may be the first to show the hypertrophy The normal markings of the bone are lost The outline is fairly regular but the surface is uneven and is broken by rather large osteophytic outgrowths These modifications are particularly common to the tibia femur fibula pelvis humerus radius ulna and clavicle The skull is thickened in some cases to two or more times its normal thickness

SYMPTOMS The onset is insidious the progress is gradual The patient may be scarcely conscious of the disorder until his attention is directed toward the deformities by his family The first symptom in some instances is pain If there is no pain medical advice is not sought until very late in the course of the disease

The pain most commonly is situated in the legs over the front of one or both tibias and is dull and felt deep in the bones It occurs in all possible grades from mere discomfort to actual paroxysms of lancinating character It is worse at the end of the day or at night and especially after long standing and unusual fatigue Pains in the skull are rare Pain may be absent throughout the course of the disease One of the most constant and severe symptoms is cramps in the muscles of the legs

EXAMINATION The muscles of the legs or arms if the bone is affected show atrophy and occasionally edema Tenderness to pressure over the muscles may be present

The most pronounced deformities are to be found in the legs No other condition except rickets in children gives such an extreme degree of curvature of the legs So great may the bowing become that the legs are crossed and walking is almost impossible The sensations and reflexes are normal

The head appears enormous and seems to rest directly on the shoulders The enlargement is confined to the calvarium which is symmetric but has a tendency to irregularities of the surface Especially great hypertrophy of the supra orbital portion of the frontal bone and the malar processes gives to the skull a massive appearance

The trunk often appears small and the thorax is compressed laterally Immense increase of the anteroposterior diameter accompanies the decrease in the lateral diameter The whole thorax is rigid and in consequence the respiration is mainly diaphragmatic The scapulas may show similar changes The spinal column becomes bowed with the most marked curve in the thoracic portion and finally complete rigidity develops The bones of the upper extremities seldom show very noticeable alterations The pelvis is very broad and massive with abnormal flaring of the iliac crests

DIAGNOSIS The diagnosis can be made in the advanced instances of the disease from the history and examination However the diagnosis is usually made by the roentgenologist from films made for causes other than bone diseases The disease thus is often discovered before there are symptoms When the disease is progressing rapidly there is increased concentration of calcium and of phosphates in the urine The serum phosphatase is greatly increased in Paget s disease The effect of immobilization results in a decrease of the alkaline phosphatase in the serum and the excretion of calcium and phosphates in the urine

The differential diagnosis of Paget s disease from malignant disease of the bone may be difficult for both the pathologist and the roentgenologist The diagnostician must be aware of these difficulties and not yield to the temptation of ordering deep roentgen therapy when the lesions are possibly those of Paget s disease

5 ***Vascular Neoplasms of Bone*** ***Benign Angioma*** (a) *Hemangioma* may form in periosteum or marrow usually in vertebrae or skull or in soft tissue where it may cause bone destruction The tumor occurs in young adults and has no sex preference It may be present particularly if situated in a vertebra in middle aged patients

Often there are no symptoms When a hemangioma is situated in a vertebra pain may be localized or may extend to the anterior part of the thorax abdomen or down the leg

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is made on the basis of histologic examination of the tissue obtained for biopsy or that removed at the time of operation The preferred treatment is irradiation when lesions are inaccessible (as in vertebral bodies) or too large to excise

(b) *Lymphangioma* was recorded by Bickel and Broders as causing almost complete destruction of the ilium from the sacro iliac joint to the acetabular region These lesions occur in the vertebrae They may occur in other bones

6 **Bone Cyst** Bone cyst occurs most frequently in the metaphyseal regions of long bones particularly in the upper ends of the humerus and the femur these account for about half of the cases Other bones involved are the distal part of the radius proximal part of the fibula distal part of the ulna and the phalanges of the hand or foot ribs pubis and ischium Latent bone cysts recognized in adult life may be present in the middle third of the shaft of the humerus and the femur Bone cysts are commoner in boys and young men than in older men or in women They may be present in middle aged patients

Often there are no symptoms When symptoms are present they consist of mild pain usually felt after prolonged use or strain of the part A pathologic fracture may be the first manifestation Trivial injury causing fracture of a long bone near a joint suggests the presence of a bone cyst

A swelling may be palpated in the upper part of the tibia or in the lower part of the radius and ulna

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is made on the basis of histologic examination of the tissue obtained for biopsy or that removed at the time of operation

Healing of the pathologic fracture may heal the cyst

7 ***Epiphyseal Chondromatous Giant Cell Tumor*** (*Codman s Tumor*) This tumor occurs in the upper end of the humerus in the region of the tuberosities in the lower part of the femur upper part of the tibia and in the metacarpal and astragalus bones more commonly in boys and young men than in older men or in women The lesion is said to arise in the epiphysis and extend into the neighboring metaphysis but it is never situated exclusively in the metaphysis

Pain occasionally swelling and limited motion in the adjacent joint are the symptoms There may be tumor at the site of pain

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is made on the basis of histologic examination of the tissue obtained for biopsy or that removed at the time of operation

Malignant Tumors Malignant bone tumors are sarcomas They arise from the diverse component parts of the bone More than half of these growths however are osteoblastic or arise from the bone itself and are therefore termed osteogenic sarcoma Those arising from the connective tissue of the bones are fibrosarcoma from the cartilage chondrosarcoma from the vessels hemangio endothelioma from the bone marrow myeloma Those arising from the reticuloendothelial cells are termed either reticulum cell sarcoma or lymphosarcoma and those from fat are liposarcoma

Osteogenic Sarcoma Osteogenic sarcomas are complex structures Only the well trained histologist can differentiate between the various types of these tumors The histologists cannot always agree among themselves on the diagnosis of the same histologic specimen

There are often stiffness and limping which may have been present from 1 to 4 years A bursitis may be the presenting symptom Acute symptoms of pain swelling and vascular compression may be due to an increased growth incident to malignant change

Inspection and palpation reveal a firm painless bulging juxta articular mass over which soft tissues move freely Pain and fluctuation may be found with bursitis

The clinical diagnosis is established by roentgenographic examination The final diagnosis is established by histologic examination of the tissue removed for biopsy or that removed at the time of operation

(b) ***Chondroma*** Chondromas occur in the small bones of the hands and feet spinal column pelvis ribs and sternum in patients from the age of 10 to 30 years

Rickets trauma differentiation of prechondral connective tissue or cartilage cells and supernumerary joint cartilages derived from prechondral connective tissue are some of the possible etiologic causes

The tumors may become large before symptoms appear However after tumor formation there may be aching pain deformity and frequently pathologic fractures The tumors may be present for years without causing symptoms

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is established by histologic examination of the tissue obtained for biopsy or that removed at the time of operation

3 ***Fibroblastic Tumor Benign Fibroma*** Benign fibroma or nonosteogenic fibroma of bone occurs in the shaft of a long bone usually with 1 or 2 inches (about 2 5 or 5 cm) of unaffected shaft between the lesion and the nearer epiphyseal plate Thes tumors may occur at any age or in either sex

Often there are no symptoms However there may be tenderness and swelling with pain and stiffness in the adjacent joint

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is established by histologic examination of the tissue obtained for biopsy or that removed at the time of operation

4 ***Giant Cell Tumors (Osteoclastoma)*** Benign giant cell tumors occur in long bones principally in the tibia femur or radius They arise in the epiphysis and progressively invade cancellous bone They occasionally present eccentrically and are called subperiosteal giant cell tumors Growth is limited by periosteum and articular cartilage They occur from the age of 10 to 40 years and have no sex preference

Trauma seems to be important There is subcortical or subperiosteal hemorrhage with subsequent atypical repair of avascular cortical bone

Pain may begin as an intermittent mild dull ache made worse by activity and diminishing or disappearing during rest Swelling is often present but it is not an early symptom When there are marked expansion and destruction of bone there is pain Movement of the adjacent joint may be limited

Palpation often elicits tumor and tenderness Pathologic fractures are common These fractures are suggested by sudden pain altered contour and increased swelling Recurrences after operation occur Malignant degeneration was present in 19 per cent of Meyerding s patients

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is made on the basis of histologic examination of the tissue obtained for biopsy or that removed at the time of operation

Bone cysts occur in the diaphysis before the adjacent epiphysis closes and are to be differentiated from benign chondroblastoma from the lesions of hyperparathyroid disease of bone which occur in the shaft are multiple and are associated with altered blood chemical findings and from central chondroma myeloma and medullary osteolytic type of osteogenic sarcoma

Accessible lesions are treated surgically There are many physicians however who advocate primary irradiation believing that better function is maintained and operation avoided

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is made on the basis of histologic examination of the tissue ob tained for biopsy or that removed at the time of operation

The prognosis is always grave only 15 per cent of patients live 5 years The usual course in the fatal instances averages about 14 to 15 months Those patients usually receive surgical treatment

2 ***Secondary Chondrosarcoma*** This cartilaginous malignant tumor originates in a benign lesion The benign tumors thus affected include cartilaginous exostosis central chondroma hereditary deforming dyschondroplasia and Paget s osteitis deformans

Secondary chondrosarcoma occurs more frequently in men between the ages of 35 and 55 years than in men of other ages or in women The most frequent sites of occurrence are the lower end of the femur and upper end of the humerus The tumors may appear about the ribs in the region of the heel or in the pelvis

There is a history of an injury a swelling which has persisted or the patient may state that there has always been a deformity in the region in question After a number of years the swelling increases in size and pain is present A pathologic fracture occurs occasionally

Aside from the presence of the tumor and perhaps some limitation of the adjacent joint the results of examination are negative

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is made on the basis of histologic examination of the tissue ob tained for biopsy or that removed at the time of operation

The life expectancy after the development of a secondary chondrosarcoma varies from 2 to 10 years The majority of patients who have been reported as cured have been treated by radical resection or amputation and not by curettage cauterization or roentgen rays

Endothelioma (*Ewing s Tumor*) The nature of this tumor is strongly suggestive of an infection The patient exhibits a generalized systemic reaction with fever and leukocytosis The histologic appearance is said to resemble that of an inflammatory lesion The disease is largely limited to the white races though it has been recorded in Negroes There is a preponderance of incidence among males

The tumor occurs most commonly in the femur and the tibia and elsewhere in the following order of frequency in the pelvis and ribs the clavicle scapula humerus radius and ulna lumbar and cervical vertebrae and the skull

The symptoms commence with pain which is variable in intensity usually intermittent becomes increasingly more severe and is commonly worse at night than during the day The lump or swelling in Ewing s tumor is present by the time there is pain and in about one fourth of the cases may be present before there is pain Fever is a common symptom and is of the intermittent spiking type The tempera ture is usually of the range between 99 and 101 F (37 2 and 38 3 C)

Spontaneous remissions occur with a decrease in the size of the tumor less pain and lack of fever only for the symptoms to return again In some cases the course is rapid The average life expectancy is less than 2 years

In a few of those who have this tumor the symptoms may have been such that the patients are operated on for incision and drainage of abscesses

The swelling is tender usually fusiform in shape and further mimics an infection with increased local heat and dilated overlying veins These tumors are further removed from the articulations than many other bone tumors and hence the dis ability is a less constant feature as an early symptom

Leukocytosis is fairly constant the leukocyte count is in the range from 10 000 to 20 000 per cubic millimeter of blood The differential blood count is not charac teristic it is not unusual in any way Anemia is not an early finding Endothelioma metastasizes to other bones to the lungs and to the lymph nodes and viscera

The clinical diagnosis is made on the basis of roentgenographic examination

Osteogenic sarcoma is essentially a disease of youth the majority of the patients being less than 30 years of age The tumor occurs more commonly in females than in males

Osteogenic sarcoma characteristically is situated in the lower extremity in the region of the knee However it may occur in the metaphyseal ends of any of the long bones It does not occur in the shaft or the epiphysis

Pain is the first symptom In the beginning it may be intermittent or transient As the tumor enlarges the pain is continuous boring persisting and worse at night than during the day

The tumor is usually ill defined and diffuse in the early stages later there is a definite mass firmly attached to the underlying bone The consistency varies greatly depending on the type of sarcoma The sclerosing osteogenic sarcoma is characteristically of bony hardness the osteolytic type is less firm in texture

As the tumor increases in size the skin becomes shiny and stretched Often there may be dilated veins in the region of the tumor and an increased surface temperature Increased heat of the tumor is often observed especially in the more rapidly growing types

Disability is produced by limitation of motion of the adjacent joint as the result of pain and tumefaction but there is no actual involvement of the joint

Leukocytosis and some fever may be present The concentration of alkaline phosphatase in the blood serum is increased

The clinical diagnosis is established by roentgenographic examination The final diagnosis is made on the basis of histologic examination of the tissue obtained for biopsy or that removed at the time of operation

The five year survival rates run consistently between 15 and 25 per cent The prognosis in general is much poorer in young children than in older persons

In general most orthopedists agree that amputation is the only treatment in attempting a cure and that all other methods are adjuncts o palliative

Fibrosarcoma This tumor arises from the nearby soft tissues or possibly from the connective tissue structures of the periosteum The femur and tibia are the commonest sites The upper extremity is involved to a less extent than the lower extremity The skull and pelvis may be involved

Fibrosarcoma occurs infrequently It tends to affect adults who are more than 30 years old

The first symptom is pain which develops slowly over months As time passes there is a swelling and then limitation of movement of the affected part The swelling is deep attached to the underlying bone and most commonly is of a semisolid consistency Occasionally but a distinctive feature when present the tumor will cross a joint and involve a second bone The lymph nodes may be enlarged These tumors metastasize by the blood stream usually to the lungs

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is established by histologic examination of the tissue obtained for biopsy or that removed at the time of operation

The prognosis of this tumor is better than that of the osteogenic sarcoma The tumor is radioresistant Surgical removal is the preferred treatment

Chondrosarcoma Some chondrosarcomas are benign while others are known to be malignant from the start and are called primary chondrosarcoma or chondroblastoma

1 **Primary Chondrosarcoma** The primary chondrosarcoma occurs in young persons less than 30 years of age with the greatest incidence in patients between the ages of 14 and 21 years The sex ratio is about the same as in the osteogenic sarcomas that is 2 males to each female

The commonest situations are the lower end of the femur upper end of the tibia or upper end of the humerus The symptoms and the findings on examination are the same as those of osteogenic sarcoma

The leukocyte count is normal True myeloma cells may be present Numerous plasma cells may be observed and if so the disease may be termed a plasma cell leukemia The platelet count is usually normal Failure of the clot to retract and difficulty in obtaining serum are often experienced by the laboratory technician Viscosity of the blood is often greatly increased The concentration of plasma protein may exceed 10 0 gm per 100 ml The increase is due mainly to the globulin fraction and the albumin globulin ratio is reversed

The serum in multiple myeloma may be anticomplementary The concentration of serum calcium is often increased (12 to 16 mg per 100 ml) but in contrast to the finding in generalized osteitis fibrosa cystica (except in the terminal stages when renal failure is extreme) the inorganic serum phosphorus is normal in amount or may even be increased The uric acid content of the blood may be increased to 25 or 30 mg per 100 ml

The urine often contains albumin and casts Bence Jones protein appears in the urine occurring in 6 of each 10 who have multiple myeloma It should not be regarded as characteristic of multiple myeloma for Bence Jones protein occasionally occurs in the urine of leukemic patients and polycythemic patients and in those who have diffuse metastasis to the ribs for instance in carcinoma of the thyroid gland The consensus is that the excretion of Bence Jones bodies in the urine is specific for some pathologic process situated in the bones that contain red marrow In the presence of very high concentrations of serum proteins the Bence Jones protein may be absent from the urine owing to an inability of the kidney to excrete it A failure to concentrate the urine and retention of nitrogen may develop

The *bone marrow* contains myeloma cells

The diagnosis is made on the basis of (1) the presence of Bence Jones bodies in the urine (2) the roentgenologist s report of myeloma or metastasis (3) increased plasma proteins (8 gm or more per 100 ml of blood) (4) difficulty encountered in making of the blood smears—rouleaux formation of the erythrocytes Despite the prolonged course the prognosis is ultimately unfavorable

Reticulum Cell Sarcoma Reticulum cell sarcoma is considered a primary bone tumor in that it arises from the parts of the reticuloendothelial system which are found in bone It may resemble histologically an ordinary reticulum cell lymphosarcoma which has metastasized to bone

This tumor has been considered to be the same as Ewing s sarcoma It occurs in an older age group than Ewing s tumor however being found in early adulthood It occurs more frequently in men than in women

Reticulum cell sarcoma occurs primarily in the metaphyseal regions of the long bones but may involve the flat bones also

Pain is the primary symptom A tumor which is tender may be present There is no fever nor is there leukocytosis

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis is made on the basis of histologic examination of the tissue obtained for biopsy or that removed at the time of operation The outlook is more favorable than in Ewing s tumor This tumor is radiosensitive

Liposarcoma The revised bone sarcoma registry of the American College of Surgeons includes liposarcoma as a malignant tumor which may be primary in bone This tumor is rare

The tumor is commonly found in the soft tissues Its existence as a primary tumor of bone is doubtful Liposarcoma of bone as well as of any other part of the body tends toward multiple lesions

The clinical diagnosis is made on the basis of roentgenographic examination The final diagnosis depends on findings on histologic examination of the tissue obtained for biopsy or that removed at the time of operation

Secondary Malignancy of Bones (Carcinoma) Metastatic malignancy to the bone is carcinoma from organs and tissue of epidermal origin such as the digestive

The final diagnosis is made on the basis of histologic examination of the tissue obtained for biopsy or that removed at the time of operation

An outstanding feature of Ewing s tumor is that the original lesion responds to roentgen therapy Despite the radiosensitivity of the primary lesion metastatic lesions nearly always develop and they are usually more resistant to treatment than the primary lesion Surgical treatment offers no better prognosis than does roentgen therapy

Multiple Myeloma Myeloma involves the bones of the skeletal trunk producing pain and resulting in pathologic fractures and anemia The disease is often distinguished by the excretion of Bence Jones protein in the urine and its accumulation in the blood However the presence of Bence Jones protein in the urine is not diagnostic for it does not occur in the urine in more than 6 of each 10 who have the disease

The etiology of multiple myeloma is unknown The disease often appears after the age of 40 years It is at least twice as frequent in men as in women

Pain tumors deformity pathologic fractures and neuralgic or neurologic symptoms are the commonest complaints and one of these may be the basis of the presenting complaint Less frequently complaints referable to anemia or abnormal bleeding or symptoms suggesting nephritis come first

The *pain* is wandering and intermittent and it is referred to the back less often to the thorax or the extremities Girdle sensations or extension down the legs may be present Generally the onset of pain is related to motion or pressure The pain may be extremely severe and may last for hours or days but intermittency and even remissions for several months are common The disease is often accidentally discovered by finding the Bence Jones protein in the urine or by evidence revealed in roentgenograms made for other diagnostic purposes

Gastrointestinal symptoms are rare but have been described as diarrhea colicky pains nausea and vomiting Hematemesis and melena have been described Epistaxis may occur and ecchymosis bleeding about the gums and lips and petechiae may be present in some of these patients

Intravascular thrombosis may occur Chronic bronchitis and pulmonary emphysema are common complications of the thoracic deformity in aging patients but are not symptomatic of multiple myeloma The liver spleen and lymph nodes are not enlarged

There may be evidences of loss of body weight The tumors are multiple and usually confined to the ribs sternum spinal column clavicles skull or to the extremities about the shoulder girdle or the pelvic girdle Generally the swellings are small the larger ones not more than a few centimeters in diameter The tumors are elastic often yielding a distinct crepitation on palpation Single palpable lesions may occur but the roentgenogram will reveal the presence of more than the one palpated Pathologic fractures are frequently present and discovered on roentgenographic examination Thoracic deformity is common

Paraplegia due to compression of the spinal cord intercostal neuralgia radiculitis diplopia anisocoria and failing vision or complete blindness due to thrombosis of the central artery of the retina as well as other neurologic syndromes may be observed with multiple myeloma as their cause

Extra-osseous tumors may be present in the testis or in the tonsil Pain in these tumors is absent if the skeleton is not involved

Anemia is of only a moderate degree if present at all Enumeration of the erythrocytes is difficult owing to clumping The clumping of the erythrocytes is related to the increased quantity of globulin in the plasma which causes a marked tendency to clumping and to rouleaux formation Clumping and rouleaux formation may prevent the making of satisfactory blood smears Blood grouping may be difficult and the serum may need to be warmed or diluted before a satisfactory test can be made Of course the sedimentation rate of the erythrocytes is increased

and postoperative convalescence It seems that irrespective of the changes in the blood or the vessel walls the essential element in the production of arterial thrombosis is slowing of the blood flow

SYMPTOMS Numbness coldness and tingling and loss of muscular power of the affected extremity are the most constant symptoms Pain is inconstant in sudden arterial occlusion It is present in only one half of those patients who have this mis fortune Pain when present is usually severe

As has been stated sudden arterial occlusions often occur during the course of a severe illness The general symptoms of the primary disease may mask those of the vascular accident If the primary disease is mild a sudden arterial occlusion may be manifested by tachycardia profuse perspiration and anxiety and pain

EXAMINATION There is absence of a pulse in the artery distal to the occlusion There are coldness a partial loss of sensation and for a short time after the occlu sion vasodilatation

After a sudden arterial occlusion there may be present gangrene or ischemic neuritis When a large artery such as the femoral is affected usually an extensive gangrene ensues which involves the leg and the foot In instances in which the lower part of the aorta is involved there are severe pain and complete ischemia and paralysis of one leg or both legs The upper extremity is less frequently affected than the lower extremity by massive occlusion of its blood supply An arterial occlusion may involve digits only When gangrene does not ensue an acute occlusion is often followed by ischemic neuritis or a state of chronic arterial insufficiency Subsequent arterial occlusions are to be expected

Ischemic neuritis is common among diabetic patients and it may merge with the so-called true diabetic neuritis Ischemic neuritis occurs most commonly when the occlusion is nearer the aorta The pain of ischemic neuritis like pretrophic pain results from a lack of blood flow to the part It is characterized by having the distribution of the peripheral nerves It may occur with or without ulceration or gangrene If there is ulceration or gangrene present the pain of ischemic neuritis extends proximally from these processes Muscular weaknesses and reflex disturb ances may follow the ischemic neuritis

DIAGNOSIS A cold pulseless extremity is diagnostically a definite entity

The prognosis as to life is poor when simple arterial thrombosis develops in the arteries of the extremities in the course of infectious diseases or congestive heart failure

Traumatic Arterial Thrombosis Penetrating wounds and severe blunt trauma may result in bruising or crushing of the wall of the artery with subsequent thrombo sis The prolonged use of a crutch may cause thrombosis of the axillary or brachial artery Cervical ribs have caused thrombosis of the subclavian artery The symptoms and findings of acute traumatic arterial thrombosis are those of acute arterial occlusion

Arteriosclerosis and Arteriosclerosis Obliterans ***Arteriosclerosis*** The etiology of arteriosclerosis is not known The consensus is that the mechanical theory and the metabolic theory are the outstanding ones in regard to the cause and development of arteriosclerosis

The advocates of the mechanical theory see the arteries change in texture as the result of continual pulsations as the years pass This change in the texture of the arteries con stitutes arteriosclerosis which reaches its most extensive development in old age The mechanical theory is not hampered by the fact that manifestation may begin in young persons for once the process is initiated the lesions tend to increase in severity and extent as the age increases and everyone grows older by the day In aged individuals who do not have significant changes in the arteries the process simply was not initiated

The metabolic theory of the development of arteriosclerosis is based on defects in lipoid metabolism It is assumed that if atheromas contain quantities of lipoid material then lipoid metabolism is at fault This theory does not seem to be hampered by the fact

tube and endocrine glands These malignant metastases are described in various parts of this book in the descriptions of the organs affected by the primary malignant focus Sarcomatous metastasis to bone is less frequent

DISEASES OF PERIPHERAL VESSELS AS THEY AFFECT THE EXTREMITIES OTHER PARTS OF THE BODY AND LOCOMOTION

Anomalies of Peripheral Blood Vessels Anomalous situations of peripheral blood vessels are often present and these seem to originate from the choice of unusual paths selected by the developing primitive vascular plexuses Other reasons for abnormally situated vessels may be the persistence of vessels which are normally obliterated and the disappearance of vessels which normally persist Incomplete development fusions and absorptions of parts usually distinct may add to the difficulty in trying to find a peripheral pulsation which ordinarily has a definite situation

Manifestations of Diseases of Peripheral Blood Vessels The presence of disturbances of the peripheral circulation is often made known by pain which is induced by exercise and relieved by rest influenced by posture localized to one digit or to a part or all of an extremity Often the pain is accompanied by color changes and both are paroxysmal in occurrence and are influenced by elevation and dependency of the part Excessive cold or warmth of extremities together with abnormal pulsations in the arteries or enlarged veins may accompany the color changes and pain There may be present ulceration gangrene scleroderma impaired nail growth excessive calluses or paronychial infections and an enlargement or a smallness of extremities

The examination of the extremities of patients who are suspected of having peripheral vascular disease can be performed quickly The examiner according to Allen Barker and Hines must answer the following questions in order to complete the examination (1) Is swelling atrophy elongation or shortening of the limbs present? (2) Is ulceration gangrene scleroderma eczema varices or thrombophlebitis present? (3) Is the temperature of the skin abnormal? (4) Is the color of the skin abnormal? Do elevation and dependency of the extremities cause unusual discoloration of the skin? (5) Are pulsations in the peripheral arteries impaired? (6) Is there auscultatory evidence of arteriovenous fistula or aneurysm? If the answer to all these questions is No all peripheral vascular disease except Raynaud s disease and erythermalgia may be excluded If the answer to any of these questions is Yes more detailed examination and analysis may be advisable

The more technical and detailed examinations comprise (1) tests of capacity of blood flow (vascular function tests) in skin as shown by vasodilatation by reflex heat artificial fever anesthesia (local general spinal and paravertebral) intradermal histamine and saline injections and reactive hyperemia (2) tests of capacity for blood flow (vascular function tests) in muscle by walking certain distances and by ergographic measurements of muscle fatigue and (3) test of the functional state of the vessels by oscillometry and roentgenologic studies especially by diodrast injection of arteries or veins Judgment much experience and common sense are needed for proper appraisal of these special tests

Arterial Thrombosis ***Simple Arterial Thrombosis*** In an occasional instance an occlusion of a peripheral artery may occur in the absence of disease of the endothelium of the artery and without a history of injury of the vessel There is no evident source of the arterial embolus In those who have such an arterial occlusion often there is a slowing of the blood flow or a change in the contents of the blood or both may be present

The blood flow is slowed during starvation severe infections and in polycythemia vera and at times in minor degrees of arteriosclerosis congestive failure in heart disease

arterial insufficiency within 30 seconds a marked degree and within more than 60 seconds an extreme degree

In arteriosclerosis obliterans there are more or less constantly the following findings a lower skin temperature of the foot or toes than of other parts of the body often scarring and shrinkage of the digits impairment of nail growth paronychias which heal slowly and ultimately may develop into gangrenous or ulcerative lesions often involving the bone of the affected digit

Ulceration and gangrene of arteriosclerosis obliterans are of the dry type and are accompanied by little or no systemic reaction When diabetes is present the lesions may be of the moist type and there may be considerable systemic reaction with rapid development of lymphangitis and often septicemia

In advanced stages of arteriosclerosis obliterans there is atrophy of the muscles of the leg absence of subcutaneous fat atrophy of the skin and often osteoporosis

Roentgenograms are of definite value in the distinction of arteriosclerosis obliterans from other types of occlusive arterial disease Roentgenograms are made of the leg the thigh and pelvis and often lateral views of the lumbar region are taken to detect calcified arteries However calcified arteries which are revealed by roentgenographic examinations may not prove or indicate occlusive vascular disease

DIAGNOSIS Intermittent claudication is almost pathognomonic of occlusive arterial disease but it is not always present

The diagnosis of arteriosclerosis obliterans is made on the basis of the absence of pulsations in one or both of the posterior tibial or popliteal arteries and unequal postural color changes in the two extremities The presence of all of the other symptoms and findings is confirmatory evidence of the disease Roentgenologic examination may reveal visible calcification in the walls of the vessels

In general arteriosclerosis obliterans is a progressive degenerative vascular disease

Thromboangiitis Obliterans (Buerger s Disease) Thromboangiitis obliterans is a segmental inflammatory obliterative disease of the arteries and veins of the extremities of young men In rare instances the disease may affect the viscera

Thromboangiitis obliterans resembles an inflammatory vascular disease The consensus is that the use of tobacco is an important etiologic factor

The lesion of thromboangiitis obliterans is a segmental inflammatory nonsuppurative panarteritis or panphlebitis with associated thrombosis without necrosis of the wall of the vessels

SYMPTOMS Intermittent claudication is the first symptom in more than 7 of every 10 who have the disease Once intermittent claudication is present it will remain present for the remainder of the life of the patient It occurs most commonly in the arch of the foot or in the calf of the leg distal to a point of demonstrable arterial occlusion Intermittent claudication is an ache sense of fatigue persistent cramp or severe aching squeezing pain It occurs only after exercise of the affected muscles and is relieved promptly by rest without change of position

Often as a first symptom some of the digits of a foot or hand may feel abnormally cold on short exposure to a cold environment Often there is an associated sensation of numbness or other paresthesias

Pretrophic or rest pain may persist for hours it is more severe at night than in the daytime and may be premonitory of ulceration or gangrene The pain of ischemic neuritis is severe paroxysmal and widespread and is associated with various paresthesias The pain of ulceration and gangrene is localized and is situated in the region adjacent to ulcers or gangrenous tissue The pain of osteoporosis and atrophy is associated with weight bearing or pressure

The lesions of superficial thrombophlebitis which occur in thromboangiitis obliterans are usually painful but the pain is not severe It persists until the lesion involutes and then gradually disappears

that although extensive arteriosclerosis increases with age there is a tendency to a decrease of concentration of blood lipoids as age advances

Disorders of lipoid metabolism manifested by lipemia often are accompanied by severe atherosclerosis The presence of diabetes mellitus is accepted as a predisposing factor to severe and premature arteriosclerosis, atherosclerosis and arteriosclerosis obliterans

Arteriosclerosis Obliterans Arteriosclerosis obliterans as it occurs in the extremities is the result of an obstruction of the blood flow through large arteries to a sufficient degree to result in ischemia of the tissues of the extremities A slowly developing occlusion may be better compensated for than the rapidly developing ones

SYMPTOMS In a slowly developing arteriosclerosis like arteriosclerosis obliterans intermittent claudication is the first symptom The pain may be present in one leg or both legs for instance in the foot in the calf or in the thigh and occasionally in the hips and lower part of the back In the majority of patients however claudication is present only in the calf muscles The pain of claudication requires fatigue for its development The pain appears while the patient is walking It is sharp and shooting darting moving rapidly lengthwise of a group of muscles The pain increases in severity with the continuation of walking Rest affords prompt relief Immediately on relief of the pain exercise may be resumed The distance that the patient is able to walk before the distress develops varies with the proximity of the arterial occlusion to the aorta The distance may vary from 20 to 25 steps to a mile or more If the pain of claudication is not yielded to there is a gradual stiffening and tightening of the muscles which when the disease is severe may reach the point that the patient is unable to use the part because of muscle spasms which are too painful to be endured After claudication there may be soreness tenderness and rather prolonged fatigue

Pain commencing while the part is at rest is rarely the first symptom This is a late symptom of advanced and severe disease Rest pain is always present if there is an acute arterial occlusion Night pain occurs in the toes the foot and the lower part of the leg It is often a persistent aching sufficient to interfere with sleep

Moderate to severe pain is associated with ulceration and gangrene Pretrophic pain like rest pain is usually worse at night than in the daytime Pretrophic pain may be so severe that it is not controlled by the administration of opiates

When there is prolonged disuse of the joints stiffness and even ankylosis may develop At least a part of this joint stiffening is secondary to the muscular weakness characteristic of insufficiency of the peripheral circulation

EXAMINATION On examination of a patient who has arteriosclerosis obliterans the findings are those of occlusive arterial disease *A decrease in the size of the* pulsations in the peripheral arteries is present The absence or impairment of pulsation in the dorsalis pedis artery is not necessarily significant Pulsations though they may be very weak *are usually present in the posterior tibial popliteal and femoral* arteries when the patient is at rest in a warm environment

The color of the feet is not changed until the arteriosclerosis obliterans is moderately advanced *When the disease is more severe the toes of one or both feet* or an entire foot are erythematous Later the reddish color changes to a bluish discoloration or pallor Immediately after an acute arterial occlusion the foot is pale and appears *shrunken*

Postural color changes such as pallor on elevation and a delay in a return of rubor on dependency are pathognomonic of occlusive arterial disease Some idea of the severity of the arterial insufficiency may be obtained as follows If the feet are elevated for 2 minutes and then placed suddenly in the dependent position failure of the color to return within 15 seconds indicates a moderate degree of

and nonvascular diseases The vascular diseases to be differentiated are Raynaud s disease acrosclerodcrma livedo reticularis acrocyanosis erythermalgia or erythromelalgia pernio ergotism acute thrombophlebitis of large veins venous insufficiency and varicose veins

The diseases which are not at least primarily vascular (nonvascular) which may require differentiation are frostbite trophic ulcers and gangrene associated with organic disease of the nervous system pronated feet metatarsalgia and other mechanical derangements associated with the flatfoot syndrome

The prognosis of thromboangitis obliterans as to the loss of life or limb is difficult to estimate

Arteriovenous Fistula The terms pulsating angioma cirsoid aneurysm arteriovenous aneurysm varicose aneurysm cavernous angioma aneurysm by anastomosis arteriovenous varix aneurysma serpentina angioma plexiforme angioma arteriale angioma arteriale serpentinum and angioma arteriale racemosum have been employed to designate or to indicate arteriovenous fistula

Arteriovenous fistula most commonly is acquired but it does occur as the result of congenital malformations

Congenital Arteriovenous Fistula Congenital arteriovenous fistula results from failure of differentiation of the common embryologic anlage into artery and vein There is a persistence of the vessels or communications which are normal in the primary anlage

Congenital arteriovenous fistulas occur anywhere in the body for instance in the bones cranium middle ear and all of the parenchymatous organs The only easily demonstrable congenital arteriovenous fistulas occur in the extremities

Symptoms of Arteriovenous Fistula in the Extremities When varicose veins appear without obvious cause over a leg or an arm of a child particularly when associated with ulceration or elongation of the limb a congenital arteriovenous fistula is suspected In some instances of congenital arteriovenous fistula the enlarged varicose tortuous veins may pulsate

Ulceration attributable to fistula may affect the hands whereas chronic venous insufficiency attributable to ordinary varices or to thrombophlebitis is singularly absent from the hands

Examination Hemangiomas (birthmarks) cavernous or diffuse port wine color or bluish red are present in somewhat more than half of those who have congenital arteriovenous fistula affecting the extremities Associated with hemangiomas may be an increased growth of hair and increased sweating

Thrills and bruits are absent in congenital arteriovenous fistula Examination to establish the presence of an arteriovenous fistula includes analysis of the composition of the blood near the fistula Arterialization of the venous blood invariably occurs For analysis the blood is withdrawn from a vein proximal to the fistula on the involved side and from a vein in a similar location on the normal side The blood from the vein of the involved extremity has an oxygen saturation which varies from 5 to 45 per cent greater than that from the normal extremity

Arteriography is performed after local anesthetization The contrast medium is ejected from the syringe into the occluded artery proximal to the site of suspected arteriovenous fistula A series of four or five arteriograms is made in rapid succession Demonstration of the opaque medium in the veins above the point of injection in the first arteriogram when the artery is closed by digital pressure is diagnostic of arteriovenous fistula The amount of filling of the venous trunks is proportional to the size of the fistula or fistulas Correct interpretation is made only by those experienced in the performance and in the interpretation of the results of arteriography

Diagnosis The diagnosis of arteriovenous fistulas of congenital origin depends on differences in oxygen saturation of the venous blood or on the results of arteri

Allen Barker and Hines separate cases of thromboangiitis obliterans into four groups on the basis of the clinical course (1) In the *nonprogressive type* after one or two episodes of arterial occlusion further episodes fail to develop and a period of circulatory compensation follows which may last for many years (2) A slowly progressive clinical course characterized by episodes of mild to moderate arterial occlusion which occur at long intervals apart Between episodes are quiescent periods of weeks to months even years when there is some improvement in the circulation as the result of development of arterial anastomoses Finally the active lesions may cease to develop and then the patient is left with a moderately to severely damaged arterial tree (3) In some patients a sudden arterial occlusion of large vessels occurs and the result is extensive gangrene If gangrene does not supervene convalescence is slow and severe arterial insufficiency persists (4) Rapidly progressive or fulminating instances of the disease with widespread gangrene necessitating amputation of the legs and perhaps some of the fingers occur but are rare

EXAMINATION The extremities may appear a normal color if the patient is in bed If the extremities are in a dependent position they are often red or cyanotic

Asymmetric postural color changes frequently are found in thromboangiitis obliterans Raynaud s phenomenon may occur in association with thromboangiitis obliterans Affected parts become abnormally blanched after a short period of elevation and on dependency after elevation there may be a delay of 5 to 60 seconds before the color returns to the skin and before the superficial veins again become visible After the return of color the part usually becomes abnormally red

The pulsations in the posterior tibial and dorsalis pedis arteries are impaired or absent in the great majority of cases of thromboangiitis obliterans Impairment or absence of radial or ulnar pulsations occurs in less than one half of those who have the disease Impairment or absence of pulsations in the dorsalis pedis artery is present as the result of anomalies of the artery in 1 of every 10 normal persons Pulsations in posterior tibial arteries may be said never to be absent as a result of congenital anomalies

Ulceration and gangrene may occur spontaneously but frequently they follow mechanical chemical or thermal trauma The sources of trauma often precipitating gangrene are injury from nails in shoes bruises nicks or cuts in the skin from trimming nails and corns or from other treatment of corns by freezing or burning and the injudicious use of strong or irritating medicaments or chemicals Nail growth is slow The nails may be thickened or thinned and deformed The digits and sometimes an entire foot appear shrunken owing to the atrophy of the muscle and absorption of fat The bones are osteoporotic on roentgenographic examination Gangrene of the hand or arm is rare

Edema of the affected extremity or extremities is a common finding A red cold swollen foot with ulceration or gangrene of one or more toes is commonly seen in thromboangiitis obliterans Superficial thrombophlebitis in association with definite occlusive arterial disease is characteristic of thromboangiitis obliterans

DIAGNOSIS A diagnosis of thromboangiitis obliterans is made when it is established that there is organic occlusion of peripheral arteries In a young man the absence or marked impairment of peripheral arterial pulsations and the presence of asymmetric temperature and postural color changes of digits of the feet or hands are diagnostic A dry gangrene of one or more digits which cannot be attributed to trauma or physical agent is almost diagnostic

Allen, Barker and Hines Sir Thomas Lewis and other authorities in the field of peripheral vascular disease warn that the presence in a limb of any one of the following conditions negatives the diagnosis of thromboangiitis obliterans (1) roentgenologically visible calcification of arteries (2) acroscleroderma or sclerodactylia (3) livedo reticularis of the legs and (4) xanthoma tuberosum

Thromboangiitis obliterans may have to be differentiated from a host of vascular

Raynaud s Disease and Phenomenon Raynaud s syndrome occurs both as a primary disease entity and as a secondary result of a number of etiologic factors

Primary Raynaud s Disease The cause of the primary Raynaud phenomenon is not known The disease is peculiarly common in girls and women and usually begins about the onset of menstruation although the first appearance may be as late as 35 to 40 years of age It is frequently observed that more than one member of the family of the patient has suffered from cold hands or feet dead fingers or other manifestations of vasospasm The importance of morbid heredity in this disease however has not been evaluated

Among the known contributing factors for precipitation of symptoms cold is important The color changes are first observed after exposure to cold Emotional and nervous upsets commonly bring on Raynaud s phenomenon although not so commonly as cold

The varying degrees of color changes in Raynaud s disease are related to transient changes in blood flow to the digits and are confined largely to the digital arterioles The changes in the digital arterioles may result from either local faults or central vasomotor factors Local faults in the digital arterioles seem to be responsible when Raynaud s phenomenon is produced only by exposure to cold Central vasomotor faults seem to be responsible when the phenomenon occurs only after emotional upsets or nervous tension

SYMPTOMS The first symptom of Raynaud s disease comprises color changes of the skin over the tips of the fingers and occasionally over the toes on exposure to cold These color changes usually begin slowly often being present in some degree for a period of years but they may be called to the patient s attention by an acute episode of pallor in one or two fingers on exposure to cold As the disease advances more fingers are affected and the extent of the disease in each finger increases until the whole digit is involved finally the backs of the hands may become affected The color changes progress successively through pallor cyanosis and rubor Pallor is present while the finger is cold Cyanosis may be present with pallor giving the lifeless hue so characteristic of the disease Rubor comes on warming the hands Pain is not a prominent symptom during the attack or in the interval between attacks

Paresthesia consisting of numbness tingling burning a feeling of tightness and a sensation of pins and needles or sticking in the fingers may occur during the attack In some cases a slight swelling of the involved fingers may occur and even persist during the interval between the attacks

In the advanced stages of Raynaud s disease the patient may become disabled from the attacks which occur at any time on exposure to a cool environment or from any emotional stress After the disease has progressed thus far the attacks may be as frequent and severe in warm weather as in cold weather

Extensive gangrene does not occur in Raynaud s disease although gangrenous ulcerations may arise on the tips of the digits and cause considerable pain and dis comfort In an occasional instance these lesions may become infected and require amputation of the distal parts of the involved finger or fingers

EXAMINATION In the interim between attacks in the mild forms of the disease no abnormalities will be observed on examination To confirm the diagnosis it may be necessary to induce the phenomenon by immersion of the extremity in cold water or by its exposure to cold air The extent nature and duration of the color changes indicate the severity of the disease

Tests for postural color changes are conducted for detection of possible occlu sive arterial disease Each peripheral artery is palpated and if questionable pulsa tions are detected in an artery help in making the decision may be gained by occlusion of the other artery or arteries to the region while palpation is continued in the questionable artery

When Raynaud s phenomenon occurs in an arm and is more marked on one side than on the other a scalenus anticus syndrome is suspected as well as a cervical rib

ography These methods may be used for the detection of arteriovenous fistulas occurring in any accessible regions of the body

It is usually impossible to correct congenital arteriovenous fistula by surgical means

Acquired Arteriovenous Fistula Acquired arteriovenous fistula of the extremities almost invariably is caused by penetrating wounds These communications may occur in any part of an extremity The passage of the arterial blood directly to the venous blood indicates an arteriovenous fistula irrespective of how devious the means of communication may be

SYMPTOMS Trauma which causes an arteriovenous fistula is followed by profuse and easily controlled hemorrhage

After trauma the occurrence of varicose veins edema stasis pigmentation ulceration and chronic indurative cellulitis round the spot where the fistula is situated indicates that the stab wound has not healed in the usual manner Ulceration and stasis ulcer in acquired arteriovenous fistula are characterized by cutaneous changes and other changes in the entire portion of the extremity distal to the fistula as contrasted with venous insufficiency from other causes in which the ulcerations are limited to the surfaces over the malleoli

Gangrene of distal portions of the extremity may occur as the result produced by shunting the arterial blood away from the capillary bed in the affected parts

If an arteriovenous fistula is acquired before the epiphyses close there is an increased length of the affected extremity

EXAMINATION On examination a thrill and a bruit continuous throughout the cardiac cycle can be felt and heard They are accentuated during systole and are diminished during diastole

The temperature of the skin of the extremity affected is increased distal to the fistula This warmth of the skin is characteristic of arteriovenous fistula

A few patients who have traumatic arteriovenous fistula are found to have enlargement of the heart A small fistula causes little if any increase in the size of the heart a fistula of moderate size produces moderate cardiac dilatation a large fistula invariably produces marked cardiac dilatation and often congestive failure When enlargement of the heart occurs in cases of arteriovenous fistula the changes which seem directly responsible are (1) a great increase in cardiac filling owing to acceleration of the velocity of blood and (2) an increase of effective venous pressure of the involved vein and anatomically associated tributary veins proximal to the fistulous connection The volume of the returning venous blood is increased because of enlargement of the artery and vein in the fistulous circuit

An increased amount of oxygen in the venous blood proximal to the fistula is indicative of arteriovenous fistula When the fistula is closed by digital pressure bradycardia (Branham s sign) is often present The systolic and diastolic blood pressures may increase simultaneously with the slowing of the heart rate Since the artery empties directly into the vein the pressure within the vein approaches that within the artery The veins may actually pulsate

The cardiac output may be increased or even doubled in the presence of a fistula of moderate size The increased cardiac size is primarily an adjustment dilatation and cardiac hypertrophy is responsible for only a small degree of the increase in the size of the heart

It has been observed by Porter and Baker that an extreme degree of cardiac dilatation can exist over a long period without cardiac failure If the fistula is large congestive cardiac failure and death may ensue if the fistula is not repaired

DIAGNOSIS The presence of a machinery like thrill and bruit which are accentuated during systole and diminished during diastole is sufficient evidence to enable one to make the diagnosis of traumatic arteriovenous fistula

The fistula should be closed surgically

of acute onset result from an arterial occlusion or an acute severe arteriospasm which is a temporary occlusion of a segment of an artery

When there is a more gradual onset after injury or operation the vasospastic changes begin weeks or even months after injury or operation In the presence of chronic irritation and infection plus the disturbed circulation bone injury follows In these cases however the osteoporosis may be largely due to inactivity or failure of normal stimulation and of use of the bone

The symptoms of posttraumatic Raynaud s phenomenon are tenderness edema and pain and cold moist slightly cyanotic skin with or without roentgenologic evidence of osteoporosis in the region of or distal to the injury The diagnosis is based on the history of trauma or operation preceding the appearance of symptoms

NEUROGENIC RAYNAUD S PHENOMENON The disturbances of the central nervous system which are commonly enumerated as being associated with functional disturbances of circulation are hemiplegia monoplegia chronic anterior poliomyelitis peripheral neuritis progressive muscular atrophy myelodysplasia syringomyelia causalgia spina bifida and cord tumors The vascular manifestations consist of persistent pallor and coldness with or without other color changes

Usually the distinction between functional vascular disturbances secondary to neurogenic disorder and manifestations of organic vascular disease is not difficult There are coldness and often color changes which are out of all proportion to the objective evidence of occlusive arterial disease

INTOXICATION In ergot poisoning both vascular and nervous symptoms are observed The vascular manifestation is gangrene that of poisoning of the central nervous system may be convulsions In some cases however both neurologic and vascular symptoms occur which produce a feeling of intense heat and cold burning pains (St Anthony s fire) and coldness and cyanosis of the extremities In the severe forms the cyanosis is replaced by gangrene which may appear suddenly The neurologic symptoms consist of severe psychic disturbances paresthesia muscular spasms and convulsions and in some cases residual anesthesia of the skin and even permanent contraction of the muscles and paralysis of the extremities may be present The neurologic manifestations are more likely to occur in persons who are malnourished than in those who are well nourished

Cases of Raynaud s phenomenon and severe vasospastic disturbance leading to gangrene from the ingestion of ergot preparations for medicinal purposes have been recorded However large doses of ergot preparations are usually tolerated without any untoward vascular manifestations

Scleroderma The terms acroscleroderma acrosclerosis and diffuse scleroderma have been used as synonyms of scleroderma Allen Barker and Hines accept the term acroscleroderma with Raynaud s phenomenon as designating a scleroderma of the extremities face and thorax which develops as a late manifestation of Raynaud s disease They restrict the meaning of the term sclerodactylia to a scleroderma which is limited to the fingers Acroscleroderma used alone implies a sclerodermic involvement of the skin of the face thorax and extremities without reference to Raynaud s phenomenon (see page 1357)

Livedo Reticularis Livedo reticularis (also called livedo racemosa livedo annularis and asphyxia reticularis) is similar in many respects to other vasospastic disorders The etiology is unknown

Since the condition is of a vasospastic nature it causes narrowing of the arterioles with an accompanying dilatation of the capillaries and venules The complaint is a persistent bluish to bluish red mottling of the skin of the legs and feet which occasionally extends to the thighs In some instances the hands and arms and the lower part of the trunk are affected The livedo never entirely disappears spontaneously It is more noticeable on exposure to cold than under other circumstances

Some patients have coldness numbness dull aching and paresthesia of the feet and legs A few have chronic ulcerations of the skin of the legs Occlusion of the

or pressure from the scalenus anticus muscle Sclerodermatous changes may be associated with Raynaud s disease or the disease may be secondary to generalized scleroderma

Diagnosis In the majority of instances Raynaud s disease can be diagnosed if there can be elicited a history of episodes of bilateral Raynaud s phenomenon precipitated by cold or emotion On examination there is an absence of primary disease such as occlusive arterial disease cervical rib or organic disease of the nervous system which could cause the phenomenon

When Raynaud s phenomenon has been restricted to one finger or to one extremity for a long time some secondary causative factor should be suspected When Raynaud s phenomenon occurs in a man it should be suspected as being due to thromboangiitis obliterans or to some other factor until conclusively proved to be otherwise If on *examination gangrene which results from Raynaud s disease is* present it will be limited to the skin Sclerodermatous changes and ulceration may complicate Raynaud s disease but these are strictly limited to the skin and no involvement of any visceral organs will be found

Patients who have Raynaud s disease are treated medically or surgically In selecting patients for surgical treatment only those who have trophic changes and scleroderma should be considered Surgical intervention may decrease their disability in cold weather

Secondary Raynaud s Phenomenon The term secondary Raynaud s phenomenon is employed to designate local changes in the peripheral circulation which *permit of changes in color of the skin resembling those of the primary* disease Allen Barker and Hines have classified the conditions or diseases in which secondary Raynaud s phenomenon may be observed as follows (1) after trauma related to occupation and following injury or surgical operation (2) neurogenic lesions cervical rib and scalenus anticus syndrome and diseases of nervous system (3) occlusive arterial disease arteriosclerosis obliterans thromboangiitis obliterans and embolism (4) intoxication for instance from heavy metals and ergot (5) miscellaneous diseases such as scleroderma lupus erythematosus and paroxysmal hemoglobinuria

Posttraumatic Raynaud s Phenomenon Two types of posttraumatic Ray*naud s phenomenon may be observed (1) occupational Raynaud s phenomenon* and (2) persistent vasospasm or Raynaud s phenomenon following injury or surgical operation

Occupational Raynaud s Phenomenon This is well illustrated by pneumatic hammer disease The rapid oscillations of the pneumatic hammer seem to sensitize the digital vessels of some workmen so that Raynaud s phenomenon occurs readily on exposure to cold There is a wide range of susceptibility to this injury Some workmen are not injured even after very long periods of using the air hammer

The episodes of Raynaud s phenomenon usually appear first in right handed men in the third fourth and fifth fingers The phenomenon does not occur during work unless *the tool is cold when in use In susceptible persons the attacks* begin within two months to a year after the worker has started to use the hammer The course of this condition is benign Often workmen are not greatly concerned about the vasospastic phenomena but accept them as being a part of their occupation

Raynaud s phenomenon may be present in pianists and typists The vasospastic discomforts involve only one or two digits Once the phenomenon is present it will continue to be precipitated by exposure to cold even if there has been a change in the occupation

Following Injury or Surgical Procedure Raynaud s phenomenon may come suddenly soon after trauma or operation or may be delayed for weeks or months The phenomenon may be associated with an osteoporosis (Sudeck s atrophy) Instances

SYMPTOMS The symptoms of obstruction of the superior vena cava are often a throbbing sensation headache vertigo and sometimes mental confusion These symptoms are exaggerated by stooping bending forward or physical exertion They are likewise increased in the recumbent position relief is often obtained while the patient is in the sitting or in the half reclining position A rapidly developing ob structive lesion produces congestion and an intense edema of the face shoulders and arms

EXAMINATION Cyanosis of the face and upper extremities and prominence engorgement and dilatation of the veins of the head neck arms and upper part of the thorax are often present

The site of obstruction in the intrathoracic veins and the veins of the upper extremities and head can be determined accurately by determining the limitations of the evidence of increased venous pressure edema and congestion If the veins of the neck and both arms are involved the obstruction is situated in the superior vena cava If a prominent plexus of large veins is seen over the sternum connecting with branches of the internal mammary vein the obstruction is probably above the level of the azygos vein If the entire superior vena caval system is obstructed large collateral veins are noted crossing the costal margin and the blood flows downward over the anterior abdominal wall

The direction of flow of blood in a dilated superficial vein is determined by stripping out the blood of a vein and compressing both ends of a segment thus freed of blood with the fingers repeat this procedure and each time remove alternately the two fingers When the finger in the direction of the blood flow is removed the vein fills slowly if at all whereas when the finger opposing the flow is removed the vein fills rapidly

DIAGNOSIS Roentgenographic examination bronchoscopic examination and if there are enlarged cervical lymph nodes biopsy of the lymph nodes in the neck or axilla are each of diagnostic value The tumors which produce obstruction of the superior vena cava are nearly always malignant

Obstruction of the Inferior Vena Cava Obstruction of the inferior vena cava usually results from thrombosis or thrombophlebitis However neoplasms aneu rysms ascites and constriction as the result of adhesive bands and scar tissue from old inflammatory processes and accidental surgical ligation cause inferior caval obstruction

Pain may be present and severe soon after the obstruction It soon subsides There are bilateral enlargement of the lower extremities with edema extensive prominence of the superficial veins of the legs and the presence of dilated and often tortuous superficial veins on the abdomen which extend well up to the thorax The direction of blood flow in these collateral veins is upward

Edema of the lumbosacral region is frequent Bilateral obstruction of the common iliac veins without obstruction of the inferior vena cava can produce similar findings The occlusion of the inferior vena cava may extend above the level of the renal veins and thus may be incompatible with life Instances of ligation of the inferior vena cava have occurred without an ensuing edema of the legs or dilated veins

Nonobstructive Diseases of Veins ***Phlebosclerosis*** Degenerative lesions of the veins are exceedingly rare Occasionally there occurs calcification of the medial coat However the so-called calcified lesions of the veins which are seen in roentgenograms of the extremities and of the pelvis are calcified thrombi (phleboliths)

Phlebosclerosis or venofibrosis according to Allen Barker and Hines occurs between the ages of 18 and 45 years The etiology of the condition has been ascribed to gout lead poisoning and syphilis and perhaps in some way it may be associated with peptic ulcer

arteries of the toes and gangrene have developed in a few who have this disease

On inspection of the extremities the reticular blotchy nature of the color changes is characteristic The diagnosis can be readily made from the appearance of the extremities Rarely does this disease cause any serious consequences though once established it is always present The disease is common in women who have fat legs

Acrocyanosis Acrocyanosis seems to represent a generally unstable vasomotor system Lewis and Landis *concluded that the fault is local* The *disease* is often a familial one which is suggestive of morbid heredity The cause of the disease is unknown

The disease affects women They complain of constant coldness and bluish discoloration of the fingers and hands which is worse during the winter than at other seasons

Examination of the peripheral arteries will not reveal any evidence of occlusive arterial disease The color changes of acrocyanosis are persistent as contrasted with the intermittent nature of the color changes in Raynaud s disease There is no history of episodes of blanching and no scleroderma or development of trophic changes ulceration and gangrene

DISEASES OF THE VEINS AND LOCOMOTION

Veins of the Leg and Foot The veins of the lower extremity are divided into superficial and deep The deep veins all have valves and there are frequent communications with the superficial veins

On the dorsum of the foot is a venous arch which unites with the medial dorsal digital vein to form the commencement of the long saphenous vein

The external or short saphenous vein begins behind the external malleolus ascends alongside the Achilles tendon thence over the gastrocnemius to empty into the popliteal vein *Its branches anastomose with those of the long saphenous on the inner side of the* leg and it communicates through the deep fascia with the deep veins It is accompanied by the external saphenous nerve

Obstructive Diseases of Veins *Venous obstruction arises from extrinsic* and intrinsic causes The possible extrinsic causes are numerous the commonest is a partial obstruction of the *iliac veins* by a gravid uterus during the last months of pregnancy Varying degrees of venous insufficiency of the limbs as well as permanent damage to the veins of the lower extremities and their valves may result from gestation

Another common cause of venous obstruction of the iliac veins is pressure from pelvic and abdominal neoplasms Neoplastic obstruction (lymphoblastomas and metastatic carcinomas) is a common cause of obstruction of the iliac veins and also of the superior and the inferior vena cava

The intrinsic cause of obstructive disease of the veins is mainly thrombophlebitis which will be considered later

SYMPTOMS The signs and symptoms of extrinsic obstruction of the iliac veins as well as the veins distal to them are enlargement of the extremity or extremities distal to the obstruction with or without edema congestion of the skin prominence of the superficial veins sometimes dilatation to the point of formation of a varix and increase of venous pressure Pain is not severe Some pain from a congestive distention in muscles *of the extremity or extremities may* occur on dependency This discomfort is relieved by elevation of the affected member

EXAMINATION In many instances the cause of the obstruction of the iliac veins is easily determined by *examination by physical means of the pelvis* and the rectum

Obstruction of the Superior Vena Cava Obstruction of the superior vena cava is a rare condition The causes of obstruction of this vessel are mediastinal tumors chronic mediastinitis *thrombophlebitis constrictive pericarditis* and congestive heart failure

Pain swelling and tenderness vary with the severity of the inflammation and the degree of congestion distal to the site of obstruction Fever tachycardia and malaise are variable among patients who apparently have the same degree of local involvement Thrombi in superficial veins are palpable

Thrombophlebitis of the saphenous systems is often secondary to infectious disease cachectic states or blood dyscrasias The multiple lesions appear as red painful tender raised linear ridges in the skin In small superficial veins the lesions may be palpated as firm cordlike segments in the course of a visible vein The inflammatory reaction undergoes involution in from 7 to 18 days but the thrombosed portion of the vein can be felt for a much longer period In some instances in the presence of arterial insufficiency necrosis of the skin occurs over the lesion (necrotizing phlebitis)

The muscular veins and deep veins of the calf are commonly affected by postoperative thrombophlebitis When this occurs there are pain in the calf and tenderness in the calf muscles Dorsiflexion of the foot will occasionally cause pain in the calf in some early and otherwise subclinical thrombosis of the muscular or deep vein of the calf (Homans sign) The presence of pain and tenderness in the calf during the immediate postoperative period may denote a thrombophlebitis of the short saphenous muscular or deep tibial veins

Thrombophlebitis of the popliteal and the femoral veins causes pain and some transient swelling and congestion of the lower part of the leg and the foot There may or may not be fever Iliofemoral thrombophlebitis is characterized by a fairly sudden onset of fever and pain present along the course of the affected vein and thence passing distalward to the foot The pain may begin in the gluteal region or in the calf The superficial veins are somewhat prominent and distended and the skin may be diffusely or locally cyanotic There is enlargement of the entire extremity

Pain and often local swelling and redness at the site of the thrombosis followed by enlargement of the extremity and often fever are *diagnostic* of thrombophlebitis A superficial thrombophlebitis involving small veins is distinguishable by the linear rather than circular shape of the lesions their smaller size lack of ulceration and comparatively rapid involution Deep thrombophlebitis of the calf may resemble myositis fibrositis and sciatic pain except that there is prominence of superficial veins

COMPLICATIONS AND SEQUELAE Chronic venous insufficiency of the limb is a serious complication of iliofemoral thrombophlebitis and saphenous thrombophlebitis

Phlebothrombosis (*Secondary Thrombophlebitis Marantic Thrombophlebitis*) In phlebothrombosis there is no evidence of antecedent disease or injury of the vein Thrombosis is first The phlebitis develops as a reaction to the thrombosis

After difficult labor cesarean section instrumental delivery and puerperal sepsis and among patients who have varicose veins or are obese phlebothrombosis is commoner than primary thrombophlebitis It has been assumed that mechanical or infectious trauma to pelvic veins may form a locus for the development of an extending thrombus in some cases However after delivery multiple thrombi occur in small veins and venules of the uterus there are changes which occur abruptly in the large dilated veins in the region of the broad ligaments there is the loss of blood during delivery and there are subsequent changes in the constituents of the blood itself

Embolism usually in the lungs is the most serious complication of postoperative venous thrombosis It is the result of phlebothrombosis and occurs in many cases in which there is no clinical evidence of thrombophlebitis Therefore it is assumed that thrombosis has occurred in some one of the peripheral veins and that either the entire thrombus became the embolus or the part of it which remained at the original site failed to produce local symptoms

The veins are small A loss of endothelium and a hyalinization of the denuded surface are followed by subintimal fibrosis

Venofibrosis rarely produces clinical symptoms

On palpation there is the sensation of hard cords which are small mobile and smooth Frequently a longitudinal groove in the skin corresponds to the location of the involved vein The diagnosis is made on the forementioned findings

Rupture A vein rarely ruptures spontaneously unless diseased Rupture is usually caused by trauma The rupture of a vein is rarely of serious importance so long as the skin is intact The hemorrhage from a ruptured vein is limited by the tissue pressure and the formation of a hematoma

Thrombophlebitis Phlebothrombosis and Pulmonary Embolism Thrombophlebitis is phlebitis in which a thrombus is present By some it is defined as partial or complete venous occlusion by an intravascular thrombus which is associated with inflammation of the wall of the vein and has a firm mural attachment Phlebothrombosis is an occlusion of a vein by an intravascular thrombus which is not associated with inflammation of the wall of the vessel is loosely attached to the wall and so can be easily detached resulting in pulmonary embolism

Thrombophlebitis Thrombophlebitis occurs (1) as a local thrombophlebitis (2) as suppurative thrombophlebitis (3) as spontaneous thrombophlebitis (4) as ischemic thrombophlebitis and (5) as nonrecurring idiopathic thrombophlebitis

In *local thrombophlebitis* there is direct injury or a pre existing disease of the wall of the vein The common example of this condition is the thrombophlebitis produced as a result of an injection of a sclerosing solution

Thrombophlebitis may originate at the site of contusions lacerations and fractures even though there is no evidence of infection of the tissues

A large number of lesions usually of the smaller veins or venules which occur as a part of inflammatory but nonsuppurative lesions are classed as *thrombophlebitis of inflammatory or toxic origin* These thrombi may be minimal or moderate in extent but are rarely extensive Examples of these lesions are the venous occlusions which occur in association with erythema nodosum tuberculosis gummas and granulomas

Suppurative thrombophlebitis may occur as the result of involvement of the wall of the vein by a severe suppurative disease In these cases the thrombus itself is highly *infected*

Spontaneous thrombophlebitis may occur in varicose veins A history of minor injuries to a varix or constriction by a tight bandage or garter may be elicited In some cases thrombophlebitis may develop in varicose veins after operations childbirth or infectious disease

In extremities in which there is arterial occlusion with the consequent *ischemia* an extensive *secondary thrombosis* of the veins is often observed The venous occlusion is usually of little importance in comparison with the arterial disease

Thrombophlebitis may develop primarily without any known injury to the vein in cases in which operation has not been performed delivery has not occurred and there has been no evidence of infectious disease noninfectious systemic disease or blood dyscrasia

Patients who have gout or those who have gouty inheritance may have thrombophlebitis migrans in which the lesions are usually bilateral and appear to migrate or jump from one vein to another

Idiopathic thrombophlebitis of the nonrecurring type often involves a large vein in a lower extremity of a patient otherwise healthy It has the same clinical features as secondary or complicating thrombophlebitis No significant predisposing factors with regard to age or sex are known (Allen Barker and Hines)

In all types of thrombophlebitis there is objective evidence of thrombosis and inflammatory reaction in the various coats of the venous wall at the site of the formation of the thrombus The red or coagulation thrombus the white or agglutination thrombus and the mixed thrombus are described The commonest type of thrombus is the mixed type

the internal malleolus or occasionally on the feet or toes if the patient has gone barefooted These ulcers often become chronically infected and odorous

Perthes test for occlusion of the deep veins of the legs is based on the physiologic actions of the skeletal muscles in the leg which influence the venous circulation toward the heart During contraction of the muscles in the leg in walking there is a compression to the deep veins resulting in a constriction and forcing the blood to pass upward toward the heart During muscular relaxation the empty and collapsed deep veins fill again with blood aspirated from the superficial veins This is known as the diastolic phase or muscular diastole

The Perthes test consists in placing a tourniquet around the thigh just above the knee constricting the superficial long saphenous vein sufficiently to prevent the reflux of venous blood and having the patient walk vigorously or flex and extend the knee repeatedly The interpretation of this test is as follows

1 If the varicosities in the leg collapse during muscular contraction the deep vein is patent permitting the blood to pass into the deep circulation through the deep communicating veins and up toward the heart This is known as a Perthes negative and permits treatment to the varicosities

2 If the varicosities do not collapse but become more distended and the patient feels pain or severe cramps in the leg during muscular contraction the blood cannot pass through the deep circulation and up to the heart because there is an occlusion or thrombosis in the deep vein—positive reaction

In the presence of certain segmental thromboses or segmental occlusions in the deep vein in the thigh the usual Perthes reaction may be erroneously negative Barone has suggested carrying out Perthes test at different levels of the leg to prevent such an error

The modified test (Barone) is carried out as follows The tourniquet is placed first below the knee above the varicosities and the patient is instructed to walk as in the Perthes test If the veins below the tourniquet collapse one repeats the test by placing the tourniquet 5 inches (about 13 cm) higher If the veins likewise collapse one repeats the test again and again each time placing the tourniquet at levels 5 inches higher up to the groin The interpretation is as follows (1) If the varicosities below the tourniquet collapse every time the patient walks with the tourniquet at different levels for example constricting the saphenous vein at different levels there is no segmental thrombus (2) If a level is reached where the veins below the tourniquet do not collapse but become more distended the tourniquet is constricting the level of the detour and preventing the blood from passing into the superficial vein through the communicating vein This is evidence of an existing thrombus in the deep vein

Diagnosis Chronic venous insufficiency is diagnosed (1) by edema of the *legs which disappears after recumbency* (2) by *dilatation and prominence* of superficial veins and (3) by a tendency to aching distress in the limb when the patient is in the erect posture relieved by recumbency in a comparatively short time particularly if the limb is elevated

The leg ulcers are differentiated from syphilitic ulcers which occur in the upper portion and often on the lateral aspects of the leg These ulcers are usually without the chronic induration and skin color changes of the varicose ulcer

DISEASES OF THE LYMPH VESSELS AND LOCOMOTION

Lymphedema Lymphedema is the manifestation of an obstruction to the central flow of lymph and the inability of the newly formed collateral channels to develop adequate valves As a result of stagnation of the lymph its protein content is increased fibroblasts proliferate and the resultant fibrosis increases the stasis Thus as the stasis of lymph increases the tissues are less viable and often inflammation is present and then more stasis ensues

There are acute and chronic lymphedemas The acute lymphedemas are commonly present during the course of acute infections

Chronic Lymphedema Homans classified the chronic lymphedemas as primary and secondary

Varicose Veins (Phlebectasia, Venous Aneurysm) Some varicose veins develop spontaneously while others develop distal to a venous obstruction

Varicose veins which develop spontaneously are the result of a hereditary weakness in the structure of the vein and its valves The veins are constitutionally poor and are unable to withstand the normal amount of stress and strain resulting from increased abdominal pressure and abdominal obesity or the opposite emaciation both of which are common as age increases Atrophic lesions of the skin such as diffuse idiopathic atrophy or acrodermatitis chronica atrophicans and extensive scars occur

Varicose veins which develop distal to a venous obstruction are a late complication of obstruction of the vena cava

Varicose veins affect both men and women

The essential demonstrable pathologic changes in varicose veins are (1) dilatation (2) elongation and tortuosity (3) loss of elasticity (4) variations in the thickness of the wall and (5) disappearance of the valves of the veins

SYMPTOMS Often extensive varicose veins of the legs may not produce any subjective symptoms When symptoms are present they consist of an increased tendency to fatigue of the leg muscles a sensation of fullness and congestion and soreness in the region of the veins

Superficially in the skin there may be an eczematoid dermatitis pigmentation paresthesias of burning pain and itching in the region of varicose veins

EXAMINATION Have the patient stand in good light with bare legs In most instances varicose veins are apparent on inspection In obese persons some varices particularly of the long saphenous vein and of other veins can be detected and examined more easily by palpation particularly with regard to their size and tortuosity

The deep veins are almost always competent unless the patient has had iliofemoral thrombophlebitis previously Iliofemoral thrombophlebitis frequently leaves the iliofemoral vein partially obstructed or with incompetent valves However in these cases compression of the superficial veins by an elastic bandage is usually well tolerated by the patient

Chronic Venous Insufficiency Chronic venous insufficiency sometimes is called venous stasis stasis edema stasis eczema stasis ulcer varicose ulcer or post phlebitic ulcer The basic cause for chronic venous insufficiency is the destruction and deformity of the small venules and capillaries to the extent that insufficient numbers are left intact to carry on without abnormal congestion

SYMPTOMS The first and often the only symptom of chronic venous insufficiency is subcutaneous edema which occurs after standing and walking The edema is above the shoe tops It will develop in the feet if the patient wears loose shoes or slippers or no shoes or slippers at all The discomfort in chronic venous insufficiency is not great A dull ache sometimes follows prolonged standing but will go within five to thirty minutes when the leg is elevated If a patient who has osteoarthritis of the knees has venous insufficiency the joint changes are often more severe In the presence of ulcers and indurative cellulitis the pain is more intense

EXAMINATION There may or may not be cyanosis of the skin The skin over the edematous regions may be irregularly covered by areas of brown pigmentation (hemosiderin) Associated with the pigmentation often there is dermatitis or eczema The eczema frequently is of a chronic scaling variety accompanied by minimal subjective manifestations It may be limited to the medial aspect over the lower third of the tibia A more diffuse eczema may be present

A hard brawny induration of the skin and subcutaneous tissues may be present If the skin is depressed over the entire circumference of the ankle and lower part of the leg or just in spots an indurated cellulitis is present Ulceration may be present and if so this is one of the common and disabling manifestations of chronic venous insufficiency These ulcerations appear just below the malleoli especially

Advanced lymphedema offers no diagnostic difficulties The brawny indurated skin and hypertrophied limb of advanced lymphedema bear little resemblance to manifestations of edema in other diseases It is only when lymphedema is not associated with changes in the appearance and texture of the skin secondary to inflammation that difficulty arises

TUMORS OF BLOOD AND LYMPH VESSELS

Benign Tumors of the Blood Vessels ***Telangiectasia*** The commonest cause of telangiectasia is exposure of the skin particularly of the face to wind and sun A less common but well known cause is excessive exposure of any part of the body to radium or roentgen rays Telangiectasia is often associated with acne rosacea xeroderma pigmentosum lupus erythematosus morphea and syphilis

Four types of localized telangiectasia are described namely spider nevus senile ectasia hereditary hemorrhagic telangiectasia and pulsating telangiectasia

A *spider nevus* (nevus araneus) appears as a raised red dot from which fine vessels radiate The lesions occur on the face neck thorax backs of the hands or other parts of the body These nevi develop spontaneously during adult life and in those who have delicate skins However they are of little clinical importance and treatment is not necessary

Senile ectasia senile vascular nevus appears as multiple small papillary lesions of a bright red or purplish color This vascular nevus does not imply the presence of any known visceral disease

Hereditary and familial hemorrhagic telangiectasia (Rendu Osler Weber syndrome) is characterized by the development of multiple telangiectatic lesions of the skin and mucous membranes The lesions occurring in the mucous membrane of the nose have a definite tendency to hemorrhage and cause frequent and often serious epistaxis by such minor trauma as sneezing coughing vomiting or strain while lifting a heavy weight The presence of these lesions on the mucosa of the oral cavity lips tongue pharynx or on the skin of the face and fingers may be the focus of excessive bleeding

The diagnosis is made from the history of bleeding from a region where telangiectasia is situated or in some instances the hemorrhage may be observed originating from the telangiectasia The history of familial involvement is suggestive The lesions may be distinguished from petechiae or small purpuric spots by their tendency to blanch on pressure from a glass slide

Pulsating telangiectasia (pulsating hemangioma) occurs on the face hands and thorax of adults The lesions are larger (2 to 5 mm in diameter) and more raised than the lesions of hereditary hemorrhagic telangiectasia

The radiating pulsating telangiectatic vessels are larger than those of spider nevi The pulsations if not obvious can be demonstrated by making light pressure by means of a glass slide over the lesion

Pulsating telangiectasia occurs in association with hepatic disease pregnancy and lymphosarcoma Severe hepatic disease was present in all of the cases reported by Williams and Snell These authors stated that telangiectasia preceded the development of the clinical signs and symptoms of hepatic disease This does not mean however that all those who have severe hepatic disease have pulsating telangiectasia or that pulsating telangiectasia is always indicative of hepatic disease

Hemangioma A hemangioma is a slowly growing benign tumor composed of newly formed blood vessels Hemangiomas are of congenital origin and are commoner in women than in men Occasionally the tumors increase in size with the onset of menses or during pregnancy

Hemangiomas are variously classified as nevus flammeus capillary cavernous diffuse racemose cirsoid and congenital arteriovenous fistulas Several of these different kinds of hemangiomas may occur in the same individual

The *primary* lymphedemas comprise the hereditary or familial the congenital the praecox and finally the simple forms The primary lymphedemas are noninflammatory and are somewhat commoner than the secondary noninflammatory lymphedemas

PRIMARY LYMPHEDEMA (Lymphedema Praecox) The onset in the majority of cases is in adolescent girls There is an accentuation during menstruation It has been postulated that the additional load thrown on the lymph vessels by rapidly developing reproductive structures induces functional incompetence or allows entrance of infection into the lymph trunks and nodes in the pelvis

Spontaneously and without any known cause the patient notices a puffiness about the foot or ankle where in most instances it stops However edema may progress up the leg and often the entire limb may be involved within a few days or weeks As the condition develops a smooth skin becomes roughened the soft edema becomes resistant to pressure tissue hypertrophy occurs and the leg becomes permanently enlarged unsightly ungainly uncomfortable, and disliked by the patient A dull heavy sensation is present Ulceration of the skin does not occur

HEREDITARY CONGENITAL LYMPHEDEMA (*Milroy s Disease*) *This condition* affects a sufficient number of blood relatives to indicate that a disturbance in the genes is responsible Its manifestations are the same as those of lymphedema praecox

The pathologic changes in congenital lymphedema are characterized by thickening and canalization of the subcutaneous tissue and the adipose tissue However in this form of lymphedema there is neither thrombosis of lymph vessels or blood vessels *nor evidence of inflammation In other forms the foregoing changes* are repeated and in addition there is atrophy of muscle nerve fibers and sweat glands and evidence of infection by leukocytic infiltration

The characteristics of congenital lymphedema are the presence at birth of a diffuse swelling involving part or all of a single extremity the disproportionate increase in the size of the involved extremity as contrasted with the uninvolved com*panion member the absence of pain ulcers and recurrent attacks of infection* and the otherwise good health of the person affected

The presence of a firm swelling which pits on pressure and is definitely reduced in size when the extremity is elevated is diagnostic In congenital neurofibromatosis the enlargement is nodular rather than diffuse and neurofibromas are not limited to a single part Congenital arteriovenous fistulas increase the length and the girth of the extremity The temperature of the skin is higher than that of the companion member The superficial veins are dilated and the oxygen content of the blood from superficial veins in the affected regions resembles that of arterial blood Congenital hypertrophy or congenital hemihypertrophy of an extremity is rare and this condition is rarely limited to a single member

SECONDARY LYMPHEDEMAS Secondary noninflammatory lymphedema includes instances of occlusion of the lymphatics as the result of malignant tumors surgical removal of lymph nodes pressure and irradiation therapy Whether lymphedema that ensues on irradiation therapy is brought about by fibrosis caused by irradiation or by metastasis of the malignant disease for which irradiation is given cannot be determined with certainty

Inflammatory Lymphedema Inflammatory lymphedema may be primary or secondary It may be a part or a result of venous insufficiency trichophytosis systemic disease or filariasis or may be due to local tissue injury or inflammation

All examples of inflammatory lymphedema exclusive of the chronic form have one feature in common namely single or recurrent attacks of acute cellulitis and lymphangitis

Pregnancy and systemic disease such as influenza typhoid fever pneumonia malaria and filariasis may lead to recurrent attacks of cellulitis and lymphangitis and result in lymphedema

anastomosis This is an end organ situated in the derma and cutaneous tissue and often beneath the nails The conglomeration of tissue acts as a control of the peripheral circulation

The glomus is compared to a thermostat in that anything which disturbs it should cause some disturbance of the heat regulation of the body When a tumor develops in a glomus the normal function is disturbed When the lesion is touched or irritated by heat or more often by cold the circulation of an entire half of the body may be disturbed by the vasomotor activation A glomus tumor may occur at any age Trauma is often associated with but probably is not the cause of glomus tumors

SYMPTOMS Pain may be present for a long time before the tumor becomes visible These lesions are usually single although multiple tumors are recorded Glomus tumors are not malignant

EXAMINATION A glomus tumor is a small purplish nodule either in the skin of an extremity or under the nail The tumor rarely exceeds a few millimeters in diameter

These tumors are so exquisitely painful that touching the lesion usually provokes a paroxysm of severe pain A satisfactory method for examination for a glomus is to ask the patient to indicate with the tip of his finger the site of maximal pain The adjoining skin is examined with the point of an ordinary steel pin The normal or surrounding skin or nail if the lesion is subungual is not unduly sensitive but the instant that slight pressure is made over the site of the glomus tumor the patient will withdraw the extremity from the hands of the examiner

Anteroposterior and lateral roentgenograms of the digit may reveal a small excavation of the phalanx from erosion caused by the tumor

DIAGNOSIS The diagnosis is established by histologic examination of the glomus tissue

The presence of a bluish or purplish lesion in the skin of an extremity and especially under the nail of a digit which when touched causes the patient acute pain is sufficient evidence to warrant suspicion of a glomus tumor The exposure of the lesion to cold will produce neuralgia like pain which seems to originate in the lesion and may spread to involve the entire extremity and may produce vasomotor changes

The tumor should be surgically removed

Malignant Tumors of the Blood Vessels ***Hemangio Endothelioma*** Hemangio-endotheliomas are malignant tumors arising from the endothelial cells of the blood vessels Hemangiosarcomas contain both fibroblastic connective tissue and vascular tissues

Hemangio endotheliomas occur in any part of the skin and subcutaneous tissue They begin as small tumors of millimeter size and increase to about 4 inches (10 cm) or more in diameter These tumors are dark red whether situated in the skin or in the mucous membranes They are soft and have a tendency to ulceration and bleeding A hemangio-endothelioma situated in the gastrointestinal tract often causes exsanguinating hemorrhages A differentiation of benign hemangiomas and hemangio endotheliomas is possible only by histologic study

One type of hemangio endothelioma (Ewing s tumor) involves the shaft of long bones particularly the shaft of the femur Two thirds of Meyerding s patients who had this tumor were boys or men less than 30 years of age The discomfort suffered by these patients was pain at the site of the lesion The correct diagnosis of these tumors can be made only by biopsy

Hemangio endotheliomas grow slowly and metastasize slowly but there are many exceptions When metastasis has occurred the prognosis is grave

The treatment of hemangio endotheliomas without metastasis consists of wide surgical excision

Nevus flammeus (port wine stain birthmark) is a form of capillary hemangioma It is a flat well demarcated purplish red discoloration of the skin which blanches easily on pressure Elevated small nodular tumors may be scattered through the lesion The lesions vary greatly in size and may be multiple They are commonest on the face less common on the trunk or extremities One of each 3 infants is born with a small nevus flammeus in the occipital region Most of these nevi gradually disappear as the children grow older Occasionally a nevus of this type is disfiguring and extends over large portions or half of the body Such an extensive lesion is usually present at birth It does not extend but enlarges as the part grows When nevus flammeus occurs on the lower extremities it may be a part of deep seated cavernous hemangiomas or multiple arteriovenous fistulas

Capillary hemangiomas (strawberry nevi capillary nevi) are circumscribed finely lobulated elevated bright red to purple red tumors which are present on the skin of the face or elsewhere at the time of birth These tumors occur singly or multiply and vary from a few millimeters to several centimeters in diameter and may attain a thickness of 1 to 2 cm They blanch incompletely on pressure

Cavernous hemangiomas (nevi cavernosi cavernomas) are tumors composed of masses of dilated thin walled vessels and are the commonest type of true hemangiomas They are present at birth They often continue to enlarge throughout life *Cavernous hemangiomas are multiple and occur in all organs of the body* The common situations are the face neck oral mucosa extremities liver bones (particularly the vertebrae) stomach and intestine In a vertebra they have to be differentiated from Paget s disease and metastatic malignant lesions On an extremity a cavernous hemangioma often involves skin subcutaneous tissue and muscle It is a localized but poorly demarcated spongy tumor with multiple bluish soft nodules resembling dilated veins just under the skin on elevation of the part the tumor shrinks but usually does not disappear Varicose veins may be associated

Diffuse hemangiomas (diffuse systemic hemangiomas) when present may involve a large part or all of one or more extremities one side of the body or a large area over a shoulder and side of the thorax

The extremity is usually enlarged and irregular The lesions resemble varicose veins of varying sizes but look queer because they are all over the extremity The affected leg feels heavy and aches after walking The lesions tend to collapse on elevation Multiple arteriovenous fistulas may coexist but their presence is difficult to prove Nevus flammeus may be associated

The presence of phleboliths may be demonstrated by roentgenograms of the extremity

Cirsoid hemangioma sometimes designated racemose hemangioma and known also as cirsoid aneurysm and congenital arteriovenous fistula is a progressive and bone eroding pulsating lesion in the region of the neck which is similar to an extensive congenital arteriovenous fistula Cavernous hemangiomas and port wine stains may be associated with multiple arteriovenous fistulas

Spontaneous involution often occurs in these angiomas therefore it is well to wait for several years before considering treatment At the very best treatment of these conditions by the plastic surgeon is not all that is to be desired

For the treatment of cavernous hemangiomas and the mixed capillary and cavernous types about the face and neck Figi stated that the implantation of two to eight 0 5 millicurie radium seeds is the preferable treatment for young children

Glomangioma or Glomus Tumor A glomus is a small conglomeration of blood vessels In the body there are various glomera for instance glomus caroticum glomus carotideum glomus choroideum and *glomus coccygeum Tumors may arise* in any of these glomera but here interest is on the cutaneous glomus digital glomus or the neuromyo arterial glomus which is the total unit making up a Sucquet Hoyer

axilla groin retroperitoneal space pelvis thoracic wall or sacral region The cystic mass is compressible and thin walled and transilluminates well Cystic hygromas become infected after respiratory infections surgical operation or radiation and when such infections occur they are always serious and may be fatal If treatment is contemplated it should be surgical removal

DISEASES OF THE MUSCLES AND LOCOMOTION

History In recording the history of a complaint referable to the muscles attention is paid to the earlier occurrence of conditions that could lead to secondary involvement of the muscles such as acute infectious disease for example acute rheumatic fever syphilis trichinosis and diseases of the nervous system

Direct or indirect trauma to skeletal structures is a common cause of muscular dysfunction The presence of pain and its situation and whether the pains or the disability appeared immediately after an injury or only some time later may be diagnostically helpful

Examination of Muscles The general musculature of the patient is inspected In some the muscles have always been small in others the muscles are voluminous The volume depends partly on heredity partly on the daily activity and most of all on the somatic type The mesomorphs are characteristically heavily muscled individuals

A completely absent muscle owing to congenital defect is rarely missed by the patient and thus no complaint is made of its absence Slight atrophy will occur whenever muscles are not used For recognition of this atrophy from disuse comparative measurements may be required

A large part of the mass of muscles lies so near the surface that individual muscles often can be easily grasped by the palpating hand others lie deep and are inaccessible On palpation the form of the muscle its tone its consistency and whether or not it is tender on pressure are observed

A relaxed muscle feels soft a contracted muscle feels firm Normally the consistency of a muscle is even throughout If firmer foci are felt they are pathologic and are often due to rupture of some muscle fibers or to abscesses hernias scars gummas or tumors Free fluid or pus within a muscle gives rise to fluctuation on palpation If the testing fingers are applied in a line at right angles to the long axis of the muscle fluctuation can often be felt in normal muscular tissue The muscle is correctly palpated by applying the palpating hand parallel to the long axis of the muscle

In making comparative measurements of the size of muscles in the arms or legs a steel flexible tape is employed and should be applied at exactly the same level on corresponding limbs thus in measuring the volume of the thigh it is well first to draw transverse lines at distances about 4 inches (10 cm) apart above the upper margin of each patella and then to measure the circumference at each of these levels It is well also to make several measurements at each level and to record the arithmetic mean A difference of less than 0 5 cm easily lies within the limits of normal variation and should be disregarded unless the difference is consistent over a considerable portion of the length of the leg or arm

Enlargement of the muscles of a part or as a whole may be due either to true hypertrophy either congenital or acquired or to so called pseudohypertrophy which is due to infiltration of fat in a dystrophic muscle

Shortening of a muscle is recognizable by a pathologic position of the limb to which it is attached thus a shortened biceps brachii causes flexion of the forearm and a shortened iliopsoas flexes the hip

It is important for instance in skin cancer to determine whether or not the process involves the muscles or the overlying fascia To determine this ascertain whether the mass becomes fixed on contraction of the muscle

Hemangiosarcoma The distinction of hemangio endotheliomas hemangio sarcomas and malignant types of spindle cell sarcomas is not always possible

Hemangiosarcomas are rare tumors which develop at any age and in any part of the body *If near the surface of the body, these tumors are soft, bluish red* and may be surrounded by smaller soft nodules Thrills are often palpated and multiple pitched bruits may be auscultated over deeply set tumors The accurate diagnosis of these tumors is made only by biopsy

The treatment of hemangiosarcomas is unsatisfactory by any means

Kaposi s Sarcoma (*Multiple Idiopathic Hemorrhagic Sarcoma Idiopathic Multiple Pigment Sarcoma*) The etiology is unknown In some instances the lesions have followed trauma or exposure to cold Approximately 9 of each 10 who have the disease are men The sarcoma is most commonly a disease of elderly people although it may affect those of any age

Characteristic of this rare disease is a multiplicity of lesions Hemorrhage occurs into the cutis and there are deposits of hemosiderin and infiltration into the layers of the cutis There are proliferations and formation of new and imperfect capillaries with cavernous spaces Metastasis occurs by way of lymph channels

The lesions often appear first on the hands or feet or in the region of the ankles but they may occur in any part of the body They vary in size from 1 mm to several centimeters in diameter and are discrete or grouped bluish red or reddish brown nodular discolorations The lesions may extend along superficial veins and often considerable edema and tenderness develop in the extremities Localized purpura and thrombosis in superficial veins are common Old lesions may appear to involute and leave atrophic pigmented scars Metastasis may occur to almost any organ in the body particularly the gastrointestinal tract liver lungs and regional retroperitoneal and mesenteric lymph nodes Some evidence indicates that the so called metastatic lesions are multiple primary foci In rare cases the earliest lesions may be in the viscera

The diagnosis is made by biopsy Kaposi s sarcoma often resembles various types of granuloma melano epitheliomas and venous insufficiency with purpura

The prognosis is poor The patients live from 2 to 10 years or longer Repeated roentgen treatment offers the only hope for prolongation of life

Tumors of the Lymph Vessels Enlargement of lymphatic vessels (lymphangiectasis) is present but not apparent in all types of lymphedema and chronic lymphatic obstruction Occasionally a dilated lymph vessel is visible appearing as *a translucent bluish or slightly pink tubule On spontaneous rupture* or aspiration a clear or slightly yellow fluid is obtained

Lymphangiomas comprise (1) simple lymphangioma (2) cavernous lymphangioma (3) lymphangioma circumscriptum and (4) cystic lymphangioma

Simple lymphangiomas are situated on skin or mucous membranes and are *benign* congenital tumors The *cavernous lymphangioma* is present at birth It grows slowly and is the commonest benign tumor of lymph vessels The tumors vary in size and may be multiple in any given area of the skin These tumors are composed of dilated lymphatic sinuses filled with lymph or mixtures of blood and lymph The fluid may congeal *become* hyalinized or calcified Cavernous lymphangiomas of the tongue lips or cheeks may cause enlargement of these parts for instance macroglossia microcheilia or macromala

Chylangiomas are cavernous lymphangiomas of the mesentery arising from the lacteals *Lymphangioma circumscriptum* occurs in eyelids conjunctivae *orbit mouth* pharynx tongue the upper extremities axillary or scapular regions and in the groin or upper part of the thigh The lesion appears as a crowded deep seated vesicle Hypertrophy of the skin may occur over older lesions giving them the appearance of warts or they may be obscured by telangiectasia

Cystic lymphangiomas (*cystic hygromas*) are present at birth They are frequently situated in the neck where they may grow upward toward the parotid gland and ear or downward to the axilla or to the mediastinum They may be present occasionally in the

presence of spina bifida poliomyelitis tuberculosis fractures neural or peroneal type of muscular atrophy or muscular dystrophies is required

Congenital Clubhand Congenital clubhand like congenital clubfoot is due to maldeveloped muscles It is observed less frequently than clubfoot

Congenital Elevation of the Scapula (Sprengel s Deformity) Abduction at the shoulder is limited since the scapula cannot be rotated Passive rotation and active rotation of the scapula are diminished or absent because of fibrous or bony attachments between the scapula and the ribs

The affected scapula is elevated and rotated so that its lower angle is nearer than normal to the spinal column and the shoulder is advanced Scoliosis and *torticollis may be associated*

The diagnosis is based on the forementioned findings Roentgenograms reveal that the scapula is shortened so that the transverse diameter is relatively increased

Congenital Contractures of the Extremities These contractures are designated also amyoplasia congenita multiple articular rigidity and arthrogryposis multiplex congenita Immobility of one or more joints is present as the result of absence or incomplete development of associated muscles

Boys and men are affected more frequently than girls and women The rigidity may affect the muscles of only one joint or the whole limb It may be bilateral and symmetric The commonest type of congenital contracture is that in which all four extremities are fixed in extension In some there may be fixation of the knees and the hips in flexion The whole limb commonly is slightly abducted

The affected limbs are often shortened and the range of movement at affected joints is limited to a few degrees because of muscles which are grossly defective or absent Congenital dislocation of the hips and other deformities may coexist Roentgenograms do not reveal any bony alterations of the joints or capsules but the muscle shadows may be very small

The diagnosis is based on the objective findings described in the foregoing paragraphs in the presence of negative results of roentgenographic examination of the joints There is little or no spontaneous improvement In severe instances surgical procedures may be helpful

HEREDOFAMILIAL DISEASES OF THE MUSCLES

The Muscular Dystrophies The term muscular dystrophy designates a disease which is characterized by progressive wasting and paralysis of the skeletal muscles The different types of dystrophies are distinguished by the age at onset the presence or absence of pseudohypertrophy and the particular muscles first affected The following forms are recognizable even if not diagnosable (1) the facioscapulohumeral type of Landouzy Dejerine (2) the juvenile scapulohumeral type of Erb (3) the pseudohypertrophic type of Duchenne and (4) the femoral type of Leyden Moebius It is abundantly clear that there is no real justification for separate descriptions of these forms

Etiology This disease may be regarded as a heredofamilial degenerative process affecting the muscles Several members of a family or their descendants are affected or the condition often can be traced through several generations Males and females are affected alike and either men or women may transmit the disease to their children It has been concluded that the mode of inheritance varies in different families and may be of at least three different types simple dominant simple recessive and sex linked recessive

Pathology The muscles may be either enlarged and firm or small and soft

No changes have ever been demonstrated in the nervous system other than the mild alterations of the anterior horn cells and motor nerve endings which may be considered secondary In recent years it has been suggested that disturbances in the sympathetic system are to blame for muscular dystrophies but there is still no satisfactory evidence to support this hypothesis

The tenderness of a muscle to pressure is tested by pressing portions of the muscle between two fingers rather than by pressing the muscle against underlying bone The tenderness elicited by the latter *method may be due to an inflamed* periosteum and not to a tender muscle

Spasm in skeletal muscle seems to be a reversible state of sustained involuntary contraction accompanied with muscular shortening and associated with electrical potential changes The diagnosis of muscle spasm is often made in various types of low back pain fractures and poliomyelitis according to Harell Mead and Mueller who used electromyographic technics and did not find muscle spasm present in these conditions The clinical diagnosis of spasm in peripheral conditions such as back ache fracture or poliomyelitis is erroneous in a large percentage of cases

Muscle spasm is differentiated from muscle spasticity, which is often caused by lesions of the upper motor neurons of the central nervous system A true spasm is characterized by sustained muscular contraction Sustained contraction or spasm is not commonly present in association with spontaneous somatic pain

CONGENITAL DISORDERS OF THE MUSCLES

The congenital malformations of muscles consist of defects and the absence of the whole or of part of a muscle

The muscular defects of greater importance are those that involve (1) the pectoralis (2) the *trapezius* (3) *the serratus anterior and* (4) *the* quadriceps femoris

These muscular defects are easily recognized by inspection and palpation of the region of the muscle and more especially by functional tests

Anomalies The origins and the insertions of muscles vary within certain recog nized limits In addition to these variations muscles may be present which have abnormal relations or attachments In an occasional individual a muscle may be present which is of no use to him but which is present in other primates as a useful muscle for example elevator of clavicle and sternalis and tail muscles Vestigial muscles are often present for example muscles for movements of the ear and scalp

Sometimes a whole or a part of a compound muscle is lacking because of *agenesis* Almost any muscle in the body may be absent or defective but there is a selective in cidence in these ageneses and defects

Some part of the pectoralis major or minor muscles may be absent the sternocostal part of the pectoralis major is that most commonly lacking The trapezius quadriceps serratus magnus semimembranous abdominals gemelli *deltoid latissimus dorsi sterno* mastoids rhomboids spinati and biceps all have been found to be absent or defective

The palmaris longus pyramidalis abdominis and plantaris muscles are inconstantly present In rare instances several members of a family may present the same peculiarity of absence or defects of muscles

Congenital Clubfoot *The deformities of the feet which are of congenital* origin are talipes equinus the plantar flexed foot talipes calcaneus the dorsally flexed foot talipes varus the inverted and adducted foot and talipes valgus the everted and abducted foot The commonest type however is talipes equinovarus in which the foot is plantar flexed and inverted In most instances the condition is bilateral

Macklin stated that this condition is inheritable and dependent on a recessive *factor* or on multiple factors and since most of these patients are males a sex linked factor is suspected The deformity often occurs in otherwise healthy babies

In the common type of talipes equinovarus the foot is inverted and flexed so that the plantar surfaces of the two feet may be brought into contact The attempt to manipulate the foot into normal position is resisted by the *ligaments and by the* shortened tibial muscles The tendons are displaced the shortened muscles are fibrotic and atrophied and the bones of the ankle and foot have come to be de formed as the result of growth in the malpositions imposed on them

The diagnosis is obvious Differentiation from acquired clubfoot caused by the

Myotonic Dystrophy (Myotonia Atrophica) This is a rare type of heredo familial degeneration of selective atrophy of muscles myotonia cataract alopecia atrophy of the sex glands and premature senility The condition is generally re garded as an independent disease although it appears to bear some relation to myotonia congenita and to the muscular dystrophies (Ford)

The onset of symptoms is usually between the ages of 20 and 35 years but it may occur in childhood The first symptoms may result from wasting of the muscles or from myotonia The muscles which are first attacked are the facial especially the orbiculares of the eyelids and the mouth the masseters and temporals the sterno cleidomastoids the extensors of the forearms vasti of the thigh and the dorsiflexors of the feet In some cases the tongue soft palate pharynx and vocal cords are affected Speech is characteristically low pitched monotonous and often nasal The hard palate is very high and narrow and the face is long and thin These patients are usually dull in mentality and are often definitely feeble minded

The myotonus is always present It is easily elicited by having the patient grip the examiner s hand with maximal force and then open the fingers as rapidly as possible It will be seen that the flexors do not relax for a second or more after the extensors begin to contract There may also be difficulty in opening the eyelids after closing them tightly cramping of the feet on attempting to run and stiffness of the jaw muscles when chewing Often repetition of the movement several times will result in disappearance of the myotonus Sudden falls may occur as a result of general myotonus provoked by an effort to run or by an emotional stimulus

Ford observes that when there is characteristic distribution of the muscular wasting the myotonus the cataracts the characteristic baldness the testicular atrophy and the appearance of premature senility distinguish this disease from the muscular dystrophies and in the presence of these findings the diagnosis is certain

The disease is incurable but may permit of survival for many years for in many cases it progresses very slowly

Myotonia Congenita of Thomsen This is a heredofamilial disease which becomes apparent very early in life and is characterized by a tendency to myotonia and often by a bulky musculature as well as by various mental disturbances The cause is unknown The disease is usually present in several members of a family and may be traced through a number of generations It affects males and females alike and may be transmitted by either sex This disease may behave as either a dominant or a recessive characteristic

The onset is in childhood It is not unusual for the symptoms to increase in severity about the age of puberty The first symptom complained of is an inability to let go quickly of an object which has been grasped firmly When the patient sud denly rises or starts to run the muscles become rigid so that the patient may be unable to take a step for some seconds or may even fall helplessly to the ground After the movement has been repeated several times the rigidity slowly wears off and the patient then is able to contract and relax the muscles normally A sudden fright a loud or an unexpected noise great anger or mental distress is almost certain to produce general rigidity in affected persons Cold weather also accentuates the myotonia

The myotonus may be found only in the legs or in the arms The facial ocular and tongue muscles may be affected Myotonia may be active or latent Latent myotonia may be elicited only by percussion by electric stimulation or by causing the patient to sneeze (use snuff) In some patients emotional reactions are the most effective stimuli

The muscles are almost always bulky even suggesting a mesomorphic somatic type or hypertrophy Strength and endurance are not diminished and it is possible for these patients to take long walks or to dance for hours

When atrophy is the main lesion of the muscle there may be resemblance in the findings to those present in lower motor neuron lesions A discussion of these relation ships is to be found under Lower Motor Neuron Lesions p 331

SYMPTOMS The insidious onset may occur at any time during childhood The parents usually complain that the child has a clumsy gait that there is difficulty going upstairs and not infrequently it is noticed that the abdomen is unduly prominent The essential symptoms include progressive weakness and wasting of certain muscle groups first the muscles of the shoulder girdle and the pelvic girdle and later the muscles of the upper arm and the thigh The wasting may or may not be preceded by pseudohypertrophy

EXAMINATION As a result of weakness of these muscle groups a number of clinical signs may be elicited which are characteristic of the dystrophies Weakness of the muscles which fix the scapula namely the serratus magnus trapezius and rhomboids results in winging of the scapula or displacement of the vertebral border of the scapula from the thoracic wall so that it stands out like a wing whenever the patient attempts to elevate the arms

As a result of weakness of the pelvic girdle and hip muscles the patient usually a child stands with the pelvis tilted forward abdomen and buttocks protuberant and a marked increase of the lumbar lordosis In walking the trunk is swung from side to side as a result of weakness of the abductors of the thighs chiefly the glutei medii The gait is not unlike that of double congenital dislocation of the hip Children who can walk fairly well on level ground experience great difficulty in going upstairs or stepping upon a chair In getting up from the floor the child turns over on the face draws up the knees and gets up on the hands and knees Next the legs are extended so that the child is resting on hands and feet with bent knees The last maneuver is to work the hands back to the legs and by grasping the legs at higher and higher levels to push the trunk erect by the arms When the process extends below the knees the dorsal flexors of the ankles are affected before the extensors so that the foot is maintained in the equinus position The knee jerks are lost early but the ankle jerks persist until late in the course of the disease Severe contractures often develop especially in the flexors of the hip and the extensors of the ankles and even deformities of the bones may occur from the long standing loss of muscle balance Sensory disturbances are absent and there is no disturbance of sphincteral control In uncomplicated cases there are never any fibrillary or fascicular twitchings

The rate of excretion of creatine is abnormal in adults In all patients who have muscular dystrophy the rate of excretion of creatinine is normal or increased

The roentgenologist may recognize a conspicuous streaking of the soft tissue shadows of affected muscles There is a delayed appearance of centers of ossification in the epiphyses of the long bones and in the bones of the hands Demineralization of the bones may be observed

DIAGNOSIS The diagnosis of muscular dystrophies is usually obvious However there may be difficulty at times in differentiating Werdnig Hoffmann muscular atrophy amyotonia congenita progressive neuromuscular atrophy amyotrophic lateral sclerosis and polyneuritis The course is usually very slow and many patients survive for 10 to 30 years after the onset of symptoms

Muscular dystrophies occurring in men and women between the ages of 40 and 55 years may be observed The women are often beginning the menopause at the time of onset In these there is no genealogic history of similar affections

The disease begins by weakness and shrinkage in the size of the affected muscles The manifestations may be of the facioscapulohumeral type or the face and the shoulder girdle may escape and the muscles of the buttocks and thighs be largely affected In rare instances there is a hyperfunctioning of the thyroid gland

In some instances the disease progresses to a fatal termination in others the symptoms may become arrested or there may be improvement or even partial recovery

Epidemic Myalgia The disease is known also as pleurodynia devil s grip and Bornholm disease The etiology is unknown The disease occurs in epidemics and affects children and young adults The disease seems to spread by contact and it is not unusual for all the children in a family to be attacked at the same time The incubation period seems to be between 2 and 4 days

Pain develops in the muscles of the back shoulders abdomen and hips Pain in the muscles of the thorax and the diaphragm is the first and often the most prominent symptom There is pain on inspiration and consequently respiration is shallow and rapid The pain is present also in the muscles of the back shoulders abdomen and hips The more acute pain lasts as a rule only 1 or 2 days but tenderness may persist somewhat longer There is only slight fever Hiccough is present in some cases

The affected muscles are tender to pressure There may be localized swellings There is mild leukocytosis and an excess of eosinophils In the presence of complications there may be found a dry pleurisy pneumonia and otitis media Orchitis is occasionally present

The diagnosis cannot be made except during an epidemic The differential diagnosis comprises the consideration of pleurisy appendicitis and dengue fever

Relapses occur in almost one fourth of all cases and second attacks are common The disease is never fatal and there are no residua

Dermatomyositis This is an acute or subacute inflammation of the muscles associated with edema and dermatitis which usually terminates fatally The involvement of muscles is widespread it consists of muscular firmness and stiffness proliferation of interstitial tissue and fatty degeneration

The disease commences with stiffness soreness and tenderness of the muscles of the extremities The trunk muscles may be involved early The skin over the affected muscles is edematous and often contains erythematous urticarial or erysipelatoid eruptions Loss of sensations of the skin is common

The course of the disorder is rapidly progressive it is fatal in 1 to 3 months and death often follows involvement of the muscles of deglutition or respiration If recovery should occur atrophy of the muscles ensues

Tuberculosis The contractile tissues of muscles rarely show tuberculous lesions so that one may speak of an immunity of muscles to tuberculosis

Parasitic Infections These include (1) trichinosis (2) echinococciasis of muscle and (3) cysticercosis of muscle These conditions have been described in connection with the respective parasites The diagnosis is established by muscle biopsy

TRAUMATIC DISEASE OF MUSCLES

Sudden and violent stretching of a muscle may produce rupture of a tendon or a muscle This causes acute pain The ruptured belly of the muscle may be displaced in the direction of the remaining attachment and when the muscle contracts the displacement becomes more evident The muscle exerts no motor effect but can be seen to contract voluntarily Rupture of the tendon of the long head of the biceps is perhaps the commonest rupture of a tendon In the same way the belly of the muscle may be torn Rupture of the plantaris muscle is apparently not very rare and may occur without violent exertion In some cases violent exertion may cause rupture of the sheath of a muscle A muscle hernia develops as a result which disappears when the muscle contracts The groove resulting from rupture of the muscle may be palpated When the muscle contracts a swelling appears on either side of the groove

The diagnosis is not difficult if the myotonia is elicited This condition rarely causes severe incapacity and does not progress Wolf has shown that quinine will abolish the myotonus of myotonia congenita

Sleep Paralysis (Dead Asleep) Sleep paralysis is frequently associated with narcolepsy and attempts by some patients to ward off an attack precipitate an attack of helplessness However sleep paralysis accompanying natural sleep is a benign condition probably resulting from a dissociation of the components of sleep It should be differentiated from familial periodic paralysis The patient is assured that although the disorder is alarming it is always of short duration and does not result in permanent paralysis or any other disease Usually just a slight touch suffices to dispel sleep paralysis

INFECTIONS OF MUSCLES

Intramuscular Abscesses Acute pyogenic infections of muscles usually result from (1) extension of an infection from the skin above or the bone below (2) the lodgment of bacteria or infected thrombi or (3) necrosis of tissue and formation of abscess from intramuscular medications

The symptoms of an intramuscular abscess are pain swelling redness and increase in local temperature Fever often is present There is often a fluctuant mass below the skin and subcutaneous tissue

The diagnosis is usually obvious, for almost all of these abscesses result from intramuscular medication The history and the findings are definite

Local Myositis Local myositis is often a complication of acute rheumatic fever and is then known as rheumatic myositis In the acute stage the inflammatory exudate in the muscle can be felt as a firm mass Later on this may be followed by the formation of scar tissue (chronic myositis) or by atrophy of the muscle

Gonorrheal myositis may be associated with gonorrheal arthritis The muscles near the affected joints are most often affected Occasionally *syphilitic myositis* is met with in the form of an infiltrating inflammation or of a gumma The so called *myositis fibrosa* is an end stage of various forms of acute myositis (infectious, traumatic parasitic) Local ossification of muscle may occur occasionally

Nodular Myositis Nodular myositis often is present in rheumatoid arthritis However nodular myositis is a comparatively common condition in a wide variety of diseases and it is of no diagnostic value in cases of rheumatoid arthritis

Myositis Fibrosa Myositis fibrosa is a subacute or chronic inflammation of the muscles occurring either locally or generally It is associated with slight constitutional symptoms and eventually ends by the replacement of the muscle fibers by connective tissue However there are many similarities between myositis fibrosa generalisata and progressive muscular dystrophy The former is essentially a degenerative and not an inflammatory lesion of the muscles It is not a separate clinical and pathologic entity but is one form of progressive muscular dystrophy

Disseminated foci are found in which the muscle fibers are undergoing hyaline and vacuolar degeneration The blood vessels are thickened

The disease begins in the thighs or calves by shortening and inelasticity of the muscles so that movements are limited in range There are no constitutional reactions such as are observed in dermatomyositis Gradually the muscles of the hips abdomen spinal column thorax and neck are involved The arms and face are usually affected very late In some cases the process is generalized and symmetric and is diagnosed *polymyositis* It may occur asymmetrically and locally

The physical examination in the beginning reveals an extra firmness of the affected muscles In advanced stages of the disease there are contractures which fix the body and extremities in a position of flexion The tendon reflexes of the affected muscles are diminished or lost No reduction of sensibility is found The diagnosis is based on the results of biopsy studies

Myasthenia gravis is characterized by weakness and by abnormal fatigue of the striated muscles which is not constantly associated with any demonstrable anatomic lesions

PATHOLOGY In some instances the thymus is found to be abnormal although the abnormality is neither constant nor specific The thymus may be enlarged it may be small degenerated and cystic or it may have undergone neoplastic changes The neoplastic change has the appearance of a lymphosarcoma No definite lesions have been demonstrated in the muscle fibers or in the nervous system

Myasthenia gravis occurs at all ages Ford recorded an infant of 18 months who had the disease There is no evidence of morbid heredity

SYMPTOMS In many cases the symptoms of myasthenia gravis are manifested without apparent cause and in many others the disease follows a respiratory infection such as influenza or bronchitis

There are abnormal fatigability and weakness and finally paralysis of the striated muscles As a rule the muscles innervated by the cranial nerves are affected first and often dysphagia may be a presenting symptom Often however diplopia is the first symptom which may accompany or be followed by the appearance of ptosis of one eyelid or both eyelids and by some limitation of ocular movement The patient may complain of being unable to look up and thus upward rotation of the eyeball is limited before any other movement is impaired The palsies of fatigue do not correspond to the distribution of any single nerve The pupils react normally In the morning the patient is refreshed and more alive than at any other time of the day As the day passes the lids droop and by evening the patient cannot hold the eyes open sufficiently to walk safely Weakness of the bulbar muscles commences shortly after the ocular muscles begin to tire The voice becomes weak and persistence in talking results in a nasal and hoarse voice

EXAMINATION Mental alertness is maintained Muscular weakness as implied by the symptoms is revealed on examination The jaws grow tired when the patient chews The face is relaxed the eyelids cannot be firmly closed and the corners of the mouth droop The neck grows weak relatively early in the course of the disease and may become so weak that the head cannot be lifted from the pillow Weakness of the bulbar and oculomotor muscles can be demonstrated before the extremities are affected The patient eventually becomes too weak to walk or stand The tongue cannot be protruded and articulation is difficult or impossible At this stage of the disease respiration becomes difficult Death may result from failure of respiration within a few hours Except for the muscular atrophy due to disuse none is present which can be determined with certainty

The muscles are atonic and soft The tendon reflexes are reduced or abolished in those who have advanced weakness In all instances of the disease the tendon reflexes are usually normal at first but on repeated elicitation the muscle becomes fatigued and finally the reflexes disappear The heart muscle and smooth muscle are not affected at all or only to a very limited extent Neither the special senses nor other modalities of sensibility are altered in any way

The abnormal excretion of creatine in the urine and the diminished excretion of creatinine are regarded as secondary effects of loss of muscle function since these findings are present in other diseases affecting the muscles Hypoglycemia too must be regarded as secondary

An enlargement of the thymus can be demonstrated in some instances on physical examination or roentgenologic examination Enlargement of the thymus has been considered etiologically important by Eaton

DIAGNOSIS A diagnosis of myasthenia gravis can be made when there is a definite abnormal weakness of the striated muscles usually beginning in the ocular or bulbar muscles and finally becoming generalized provided there have been remissions and exacerbations of the weakness The abnormal weakness of the muscles is

The history and the findings are diagnostically conclusive

As the result of direct trauma all or a part of a muscle may be ruptured For instance a part of the quadriceps femoris muscle is often ruptured from the trauma of falling forward and astride the *steering wheel of an automobile when the over* turning car comes to a sudden stop

HERNIA OF MUSCLE

Hernia of a muscle due to a defect in the aponeurosis that permits a mass of muscle to bulge through it on contraction may follow trauma but it often results from congenital fascial defects

Hernia of a muscle is diagnosed by observing and palpating the muscle during voluntary contractions

Fascial Hernias of the Legs Fascial hernias of the legs occur unilaterally and bilaterally in the muscles of persons whose legs have been exposed to severe chronic strain such as athletes *skiers mountain climbers foresters and foot soldiers who* have exercised in hilly or mountainous country

CONTRACTURE OF MUSCLES DUE TO CIRCULATORY DISTURBANCE

Volkmann s Ischemic Contracture The most serious and one of the com monest types of traumatic injury from ischemia is Volkmann s ischemic contracture However comparable contracture may occur as the result of circulatory disturb ances in compressed injured muscle in any part of the body

It is generally accepted that circulatory disturbances are responsible for ischemic contractures In Volkmann s contracture the application of tight splints is usually held to blame However hemorrhage and edema under the fascia in the antecubital space could compress and obstruct the blood vessels there and produce the contracture even if a cast to the arm had been originally correctly applied The application of a tourniquet or occlusion of the large vessels of the arm may cause the contracture

In Volkmann s contracture there is at first edema followed by necrosis of the muscle fibers The fibers are gradually broken down and replaced by connective tissue The muscle becomes a mass of scar tissue which is adherent to the skin the tendons and bones

Volkmann s contracture is found almost exclusively in the muscles of the fore arm which flex the fingers and wrist It follows severe injury to the region of the elbow joint such as fracture of the lower end of the humerus or of the bones of the forearm Application of casts which are too tight a cast made too tight after appli cation by hemorrhage under the fascia and occasionally application of a tourniquet are the common causes of ischemic contractures Ischemic contracture is very pain ful and is characterized by pronounced swelling and cyanosis of the fingers

On examination the fingers are cold and blue Later the skin is drawn tight and is adherent to the subcutaneous tissues and shrunken muscles The fingers and perhaps the wrist are flexed so strongly at the distal joints that the nails may press into the palm *but the proximal joints are extended The thumb may or may not be* affected The atrophy and induration are mainly of the extrinsic and intrinsic flexors of the hand and fingers

The diagnosis *in the fully developed condition is obvious If the condition can* be recognized before irreparable damage is done the splints should be removed at once If the circulation does not improve satisfactorily an incision should be made into the antecubital space and the blood clot removed *The prognosis is bad*

TOXIC AND METABOLIC DISORDERS OF MUSCLES

Myasthenia Gravis Synonyms for myasthenia gravis are asthenic bulbar palsy bulbar palsy without anatomic basis myasthenia gravis pseudoparalytica and Gold flam s disease

The Tic Diseases A tic is a co ordinated muscular movement that has become imperative a function il disorder The movement may correspond to that of a reflex defensive or expressive type The tic is response to an irresistible impulse to move ment which makes the patient uncomfortable until the action is performed It is brief may be violent repetitious and limited to a single movement or it may be characterized by a multiple series of movements

Isolated Tics These tics are commonly present in the facial muscles and repre sent specific acts such as sucking licking and biting Scratching and rubbing par ticularly of the nose or face may represent a tic

Generalized Tic The disease commences as a form of facial tic—blepharo spasm or *distortion of the mouth—or with a head nodding tic Later stereotyped* tics in the extremities appear Practices without reason or emotional urge are in dulged in such as grabbing of the nose pulling the chin or throwing the head to the side hand-clapping and foot stamping In some instances the tics seem to be part of a psychasthenic state in others of a dementia praecox A generalized tic once manifested does not stop

Generalized tic is distinguished from Sydenham s chorea from hysteria and from double athetosis These patients are usually schizophrenics

Paramyoclonus Multiplex The disease is known also as myoclonia poly-clonia and Friedreich s myoclonus The clonic contractions cause short quick jerks of the extremities not involving synergistic muscle groups and varying in number from a few to 100 per minute The muscles often involved are the brachioradialis biceps trapezius quadriceps femoris and semitendinosus The contractions disap pear in sleep The deep reflexes are exaggerated

The disease must be differentiated from the tic diseases hysteria petit mal and chorea chronica

Writers Cramp Persons of various occupations are prone to a disturbance of special skilled co ordinated muscular movements used in performance of their work

Writers cramp develops in psychoneurotic patients The disturbance commences with an abnormal fatigability on writing later there is a feeling of cramp in the muscles The writing becomes abnormally irregular almost illegible especially when the writer is under observation Results of physical examination of the nervous system may be negative except for the presence of psychoneurotic signs

Writers cramp is distinguished from agraphia paralysis agitans hemiplegia tabetic ataxia beginning in the upper extremities familial and acquired intention tremor and multiple sclerosis

TUMORS OF THE MUSCLES

Tumors of muscles may be benign or malignant The benign tumors include lipomas fibromas and osteomas The malignant tumors include as primary tumors angiomas and sarcomas and sometimes as a secondary tumor carcinoma Tumors of muscles should not be confused with myositic indurations tuberculosis gumma or parasitic invasions

Desmoid Tumors of Muscles (Fibromas) A desmoid tumor may be defined as a very hard tough fibroma Multiple desmoid tumors may occur in striated muscles especially in the anterior abdominal wall However these tumors either singly or multiply occur elsewhere in the body

That these tumors are due to trauma (accidental operative or the physiologic trauma) combined with an unknown individual predisposing factor appears to be the most logical theory of origin These tumors do not undergo metastasis nor do they endanger life however they do tend to recur locally *Diagnosis is made by* finding a fibrous tumor in a muscle the tumor is fixed by contraction of that muscle Biopsy is indispensable for diagnosis

revealed by an increase of symptoms in the evenings and by fatigue in repetition of movements of affected muscles An improvement in the symptoms which follows the injection of neostigmine (prostigmin) is of diagnostic value for such always occurs in myasthenia gravis and never so dramatically is *improvement witnessed* in other conditions Myasthenia gravis is rare in childhood and in this period the diagnosis must be made with caution

Quinine sulfate will precipitate the symptoms of myasthenia gravis Curare in *very small doses* is used as a test for myasthenia gravis Patients who have this disease are very sensitive to curare and for this reason this drug should not be used by those who have not had supervised experience in its use When curare is used as a test for myasthenia gravis not more than one twentieth of the curarizing dose is employed The test is positive if great muscular weakness is produced by the administration of curare

There is no curative treatment at present Prostigmin (5 to 15 mg doses) is of great value but its effect is purely symptomatic The effect of each dose lasts about two hours

In some instances the symptoms advance rapidly and death supervenes within a few months In others in which symptoms are definite there is a tendency for them to remain stationary for a long period In some instances there are recurrent exacerbations with complete remissions between

OSSIFICATION IN MUSCLES

Hematomas and granulation tissue resulting from inflammatory processes in muscles may eventually become ossified Bits of periosteum torn from the bones as a result of trauma give rise to formation of new bone Long continued trauma of a milder type may also lead to bony deposits in the muscles or tendons For example plaques of bone are found in the adductors of the thighs in individuals whose occupation requires daily horseback riding Extensive bony processes sometimes appear in the muscles of the lower extremities in severe spastic paraplegias of long duration These conditions are localized and do not progress beyond local limits

Progressive Myositis Ossificans Progressive myositis ossificans is a generalized ossification of the skeletal muscles which is dependent on some inborn defect The etiology is unknown (see also page 1535)

In the beginning there is muscle edema and often there are hemorrhages The onset begins before the tenth year There is a local firm nontender swelling situated in the muscles of the trunk or back of the neck The skin may be slightly red and tender The swelling slowly disappears leaving a firm small region in the muscle which eventually becomes bony In time successive swellings develop leaving a bony deposit

When the disease is fully developed it can scarcely be confused with any other condition The roentgenographic appearances are characteristic

In those who survive long enough the disease progresses sufficiently to produce invalidism and death from inanition or fixation of the respiratory apparatus

MUSCULAR DISORDERS SECONDARY TO DISTURBANCES OF INNERVATION

These disorders include (1) the muscular atrophies due to lesions of the lower motor neurons and peripheral nerves (2) the muscular atrophies due to disuse and (3) hyperkinetic disturbances

The muscular atrophies due to lesions of the lower motor neurons and peripheral nerves are described on pages 330 and 331 The atrophies of muscles from disuse occur when a joint is mobilized as by a plaster cast or ankylosed

The hyperkinesias comprise *the tic diseases choreas and the occupational neuroses*

wasting small groups of muscle fibers show irregular twitchings known as fibrillary twitchings or fasciculation The degenerating muscle is usually extremely tender to pressure and may be painful and subject to cramps

During this phase of degeneration in the muscle it may show what is known as the reaction of degeneration The electric testing of muscular reactions is of no value unless performed by an experienced hand and it is comparatively rare for this testing to provide diagnostic information that cannot be obtained by other means Of greater diagnostic importance is the fact that apparatus for such testing is not usually available in practice

It is immediately apparent that *lesions of the lower motor neuron* and the neural or *peroneal muscular atrophy* have symptomatically much in common A neural or peroneal atrophy is characterized by a slowly progressive palsy with flaccidity of muscles A *hypertrophic interstitial neuritis* causes the same symptoms but can be recognized by the palpable thickening of the peripheral nerve trunks In either disease more than one member of a family may be affected

In the descriptions of the diseases of the muscles it was recorded that the *myopathies* are characterized by chronic and progressive wasting of the muscles and frequently appear in several siblings The muscles of the pelvic girdle and the shoulder girdle are selected by this process and the muscles of the proximal parts of the extremities are also affected early in the course of the disease

Muscular atrophies and flaccid paralysis are due either to a lower motor neuron lesion or to lesions in the nerve or the muscles When the lesion is situated in the muscle it is comparable to the lesions present in the muscular dystrophies The muscles are soft waste away and throughout the illness lack tone There is absence or diminution of the tendon reflexes The plantar response is flexion When the lesion is in the lower motor neuron tests of electric excitability reveal loss of faradic reaction and a slow galvanic response that is the reaction of degeneration When the lesion is in the muscles there is only a quantitative reduction of irritability Atrophy of the muscles must be distinguished from defective development When the lesion is situated in a peripheral nerve trunk there are muscular atrophy and sensory changes in the skin

Among the *congenital conditions* giving rise to a generalized chronic congenital weakness and loss of tone of the muscles are amyotonia congenita and atonic cerebral diplegia Both of these diseases are uncommon

Among the conditions causing flaccid paralysis at birth are rupture or stretching of the brachial plexus during birth which frequently causes paralysis of the fifth and sixth roots of the plexus with loss of abduction and external rotation at the shoulder of flexion at the elbow and of supination of the forearm Injury to the upper roots of the brachial plexus during birth may cause paralysis of the diaphragm on one or both sides

Acute flaccid palsies in childhood and adulthood often are due to poliomyelitis trauma peripheral neuritis and injury to muscles as occurs in Volkmann s ischemic contracture Poliomyelitis is described immediately subsequent to this discussion

Because of the high incidence of local palsies with atrophy as the result of injury to the spinal nerves these are always considered in any diagnosis when these findings are present The nerve may be lacerated by fragments of bone injured by pressure of a tight splint or compressed by formation of scar tissue or callus Falls which depress the shoulder may cause brachial plexus palsy

Acute Anterior Poliomyelitis (*Infantile Paralysis Heine Medin Disease*) Poliomyelitis is an acute infectious disease that is caused by the smallest in size of the filtrable viruses which has sufficiently distinct properties to permit its identification with much certainty

There are three immunologically distinct types of the poliomyelitis virus type 1 (Brunhilde) type 2 (Lansing) and type 3 (Leon) These types are so sharply

Venous Angiomas of Skeletal Muscle In some patients angiomas of the muscles are true tumors They may grow and exhibit the characteristics of a malig nant neoplasm In others angiomas are malformations masses of swollen blood vessels which are of congenital origin or clearly related to injury Their course is variable many remain stationary some disappear spontaneously The angiomas of congenital origin do not produce symptoms until late childhood or early adult life

Venous angiomas of skeletal muscle occur with about equal frequency in the two sexes In most cases the malformation is first observed during childhood or early adult life

The symptoms commence with pain and swelling related to muscular movement The tumor and thus the symptoms may occur in any striated muscle although the most frequently involved muscle seems to be the quadriceps extensor Venous angiomas vary in diameter from less than 1 inch (1 to 2 cm) up to 4 or 5 inches (10 to 12 cm) or more

Paresthesias numbness and periodic or constant swelling are commonly present There is occasionally fixation of the overlying skin which exhibits some degree of bluish discoloration The mass is soft and movable rarely is it firm and fixed Seldom are pulsations or bruits to be made out unless there are free arteriovenous com munications

In addition to the clinical examination there are three procedures which are diagnostically helpful (1) exploratory puncture with aspiration of normal blood (2) the intravenous injection of radiopaque solution and (3) roentgenologic ex amination The diagnosis is based on the history and the findings on examination

LOCOMOTION AND THE NERVOUS SYSTEM

In man the basic structures subserving locomotion consisting of bones joints muscles and nerves have not changed greatly since he began to walk on his hind legs The great change has come about through freeing the front quarters of their main responsibility in locomotion The front quarters having been emancipated from responsible locomotor functions come more under the influence and guidance of the cerebral cortex than do the front quarters of the animals who use them solely for locomotion It therefore has been necessary to consider the organization of the motor nervous system as such after the description of the diseases of locomotion as they occur in bones joints muscles and peripheral blood vessels

The neuron concept of the development and structure of nerves and nerve fibers re pudiates the idea that nerve fibers develop from cell chains or a syncytium and holds (1) that all axons and dendrites are true outgrowths from nerve cells and (2) that each neuron remains throughout life a discrete structural and functional unit Critical cytologic observations favor discontinuity where process and cells of different neurons come into functional contact Collateral evidence is afforded by the behavior of neurons after severance or injury the fibers distal to the point of section (and thus isolated from their cell bodies) degenerate whereas the central stumps live and regenerate

THE MOTOR NERVOUS SYSTEM

On the basis of structure and function the motor nervous system is divided into five components namely (1) the lower motor neuron (2) the upper motor (pyramidal) neuron (3) an extrapyramidal system of neurons (4) the cerebellar system and (5) the frontal cortex (eupraxic functions) immediately anterior to the motor cortex

The Lower Motor Neurons and Locomotion Any lesion which interrupts the lower motor neuron deprives a muscle of its nerve supply and causes it to be come flaccid and completely paralyzed to lose its tendon reflex and to waste Fibrous changes take place in its structure it shortens and undergoes contracture In cases in which the interruption of nerve supply is incomplete during the active process of

are no data to support a contention that dietary deficiencies predispose to polio myelitis On the contrary it often seems that the victim of poliomyelitis is well nourished perhaps even better nourished and developed than the children who do not get the disease

PATHOLOGY Focal areas of injury are present in the pons brain stem motor cortex and meninges Injuries have been found in the dorsal root ganglion There are irritation and hyperplasia in mesenteric and enteric lymphatic tissue which are especially con spicuous in children

In the spinal cord the lesions are most prominent and extensive in the cervical and lumbar enlargements especially in gray matter of the anterior horns It is the destruction of ganglion cells in these areas that is responsible for the clinical expression of the disease Pathologically the disease is associated with destructive lesions in the gray matter of the spinal cord and brain the white tracts being largely spared

Depending on the severity of the disease process or the situation of the main involve ment in the spinal cord several types of poliomyelitis have been designated In the *abortive* type there are constitutional symptoms without symptoms of invasion of the central nervous system In the *nonparalytic* or *meningeal* type there are constitutional symptoms of the abortive type plus fever drowsiness and signs of meningeal irritation and an increase of cells in the spinal fluid No paralysis occurs in these cases The *spinal* type embraces the classic forms of the disease When symptoms indicate involvement of the medulla pons or midbrain occurring alone or combined with spinal symptoms the disease is designated the *brain stem* type

SYMPTOMS In most instances of poliomyelitis the first symptoms are referable to the gastrointestinal and respiratory systems singly or combined There are fever headache catarrhal inflammation of the respiratory passages and often diarrhea The temperature reaches its peak in the afternoon 100 to 103 F (37 7 to 39 4 C) and shows a tendency to drop in the morning In from 1 to 3 days symptoms referable to the central nervous system appear In a few cases a remission follows the initial symptoms Then the temperature rises again and paralysis may or may not supervene An occasional case of poliomyelitis is observed without initial symptoms and is characterized by an abrupt appearance of paralysis

Headache is the most distinctive initial complaint in poliomyelitis This is usually of a severe generalized unrelenting type which is a rare symptom in the illnesses of childhood In addition to headache and fever there are nausea vomiting anorexia stiff neck stiff back painful extremities and general malaise with listless ness The muscular pains may be severe Sore throat may be an early complaint The absence of inflammation of the throat in patients who complain of sore throat sug gests the possibility of a painful throat of neuritic origin The complaint of sore throat occurs more frequently in those in whom bulbar paralysis develops than in other patients There may be drowsiness and apathy Irritability alternating with drowsiness is common Sometimes the drowsiness deepens into stupor or in fatal instances into coma Within a short time signs and symptoms indicative of involve-ment of the segmental motor apparatus develop The fever lasts for 4 or 5 days and falls by lysis although it may persist for several weeks The prostration and drowsi ness may continue for several days after the fever has ceased

In the cerebral type there is a profound stupor both with and without spinal palsies Included here also is hemiplegia the type with an acute febrile onset without spinal palsies (Strumpell s polioencephalitis)

The paralysis remains unchanged for several weeks before improvement com mences The major part of the improvement may be expected within six months but appreciable returns of tone and muscle power may be observed after the end of the first year There is atrophy of the affected muscles on the permanency and extent of which depend the spinal lesions The paralysis of groups of muscles imposes the additional handicap of lack of muscle balance The soft bones of childhood are unable to cope with a lack of balance to carry the body weight and the ankles

defined that convalescent serum from one type will not protect against the others and also that the convalescent monkeys which are immune to reinoculation with one type can be reinfected with another type Many observers believe that in the human being the virus of poliomyelitis is distributed predominantly in only two systems the nervous system and the alimentary tract

In human infections probably the virus enters the body by way of the mouth and establishes itself throughout the alimentary tract where it proceeds to multiply There is suggestive evidence that the walls of the pharynx and small intestine are among the chief sites of virus multiplication while the virus is in the alimentary tract and that the feces are the chief means of virus elimination The virus may begin its invasion of the central nervous system along the cranial nerves that supply the upper part of the digestive tract and thus pass into the medulla It may enter by way of the parasympathetic system along the vagus from the middle part of the digestive tract into the spinal cord It could pass along the visceral afferent fibers through the spinal ganglia or along the visceral efferent fibers from the intestine by way of the abdominal sympathetic ganglia

Poliomyelitis is cosmopolitan in distribution It occurs in epidemic sporadic and endemic forms No race is immune In the United States it is most prevalent in Vermont New York New Jersey Nevada and Minnesota More than three fourths of the sporadic cases occur between June and October and all the epidemics have prevailed during the warm months of the year

The *disease is contagious* being transmitted directly by droplet infection by healthy carriers of the virus by those who are in the prodromal stage or by those acutely ill with the disease

Children between the ages of 2 and 4 years are most susceptible to poliomyelitis the disease is unusual in early infancy and in adult life In no age group however is there full immunity Blood serum of convalescents and of many adults who are not aware of ever having had poliomyelitis contains antibodies thus indicating that the morbidity rate is much higher than is recognized from the clinical evidence of having had the disease A high degree of immunity is conferred by an attack of poliomyelitis since second attacks are exceedingly rare

The incubation period varies between 7 and 14 days and often from 1 to 6 days In Casey s experience the incubation period (calculated from exposure to onset of prodromal period) in 37 instances of epidemic human poliomyelitis varied from 5 to 35 days and averaged 12 2 ± 1 1 days

Casey Fishbein and Bundesen observed that *multiple cases of poliomyelitis in the family* are the rule rather than the exception when there are other children from about 2 to 9 years of age in the home The disease is less infectious in the older groups In many instances of poliomyelitis the disease was so mild that a physician was not consulted Once the disease is present in a neighborhood there is no evi dence that flies and other insects play a major role in the spread of the disease

Cunning presented a statistical report which completed a four year nationwide survey conducted by the American Laryngological Rhinological and Otological Society This study was based on 36 678 cases of poliomyelitis and 96 379 cases in which *tonsillectomy* had been performed It failed to reveal any causal relationship between poliomyelitis and tonsillectomy The studies did not indicate that tonsil lectomy should be indefinitely postponed in the summer months simply because poliomyelitis is prevalent during this season The author did not however advise operation during any epidemic regardless of its nature

Clinicians generally agree that a deficiency in vitamins or other essential food elements usually results in a lowered natural resistance to bacterial infections There is reason to believe that this hypo immunity is largely due to a reduction in phago cytic functions In contrast with this general agreement the effect of similar nutri tional deficiencies on antiviral resistance is still controversial At present there

are affected in addition to the diaphragm respiratory failure and death ensue Intercostal paralysis is recognized by failure of the thorax to expand and by retraction of the lower ribs during inspiration

Involvement of the muscles which aid in support of the spinal column usually is not detectable until some time after the acute stages of the disease have passed and severe spinal deformities develop

The affected extremities are cold mottled with a purplish blue color and slightly swollen

The brain stem type of poliomyelitis selects the motor nuclei of the cranial nerves As a rule however the cranial nerve paralyses do not persist Third and fourth nerve paralyses are common The fifth and the twelfth nerves are usually spared The facial nerve is usually spared The sixth nerve is the one commonly affected in ocular palsy Paralysis or weakness of the soft palate the pharynx and the vocal cords and irregularities of pulse and respiration are present in tenth nerve involvement The voice is hoarse and swallowing is difficult

There is leukocytosis the concentration of leukocytes ranging from 15 000 to 30 000 per cubic millimeter of blood beginning in the prodromal stage and persisting for 6 or more weeks The lymphocytes may be diminished and thus occasionally the total leukocyte count is normal owing to this reduction of lymphocytes and increase in other white cells

During the first week the cell count of the spinal fluid is often between 50 and 100 or more per cubic millimeter Lymphocytes predominate in the spinal fluid throughout the course of the illness In 2 weeks the cell count returns to normal values The globulin content during the first week is normal Thereafter its value increases and the increase may persist for several weeks to more than a month The content of sugar and chloride is within normal limits

The spinal fluid is never purulent and yellowish in poliomyelitis The only time that there is an increased polymorphonuclear leukocyte count in poliomyelitis is during the early stages of the disease prior to the time when paralysis is present If the cells are predominantly leukocytes the probabilities are that bacteria cause the condition and they can be identified on a stained smear

When there are mixed types of cells and the proportion of lymphocytes is greater than that of leukocytes either the patient is recovering from an acute infection which might have been pyogenic in origin commonly an irritation of the meninges or some disease is developing which ordinarily causes a lymphocytic reaction A second puncture to determine the further progress of the disease is indicated If the ratio becomes more lymphocytic it is recognized that probably the disease is one causing lymphocytic reaction If the cells are predominantly lymphocytes it is known that the disease is an encephalitis of some character

The amount of protein present in the spinal fluid gives a clue to the severity of the involvement

Diagnosis The presence of fever signs of meningeal irritation flaccid palsies and the changes in the spinal fluid are diagnostically definite

During the summer any patient from 3 to 10 years of age who has difficulty in speaking difficulty in clearly enunciating syllables sounds words and especially consonants or who has any difficulty in swallowing solid foods or liquids should be suspected of having poliomyelitis

Examination of the spinal fluid is the important diagnostic aid in the preparalytic stages of the disease During the progress of an epidemic the nonparalytic form of the disease may be suspected or diagnosed but when this form occurs sporadically it cannot be diagnosed

In influenza there may be a stiff neck and the gastrointestinal symptoms may be the same as those in early poliomyelitis However the neurologic and spinal fluid findings are negative

knees hips and back are forced to become deformed Paralysis of the intercostal muscles may permit gross deformities of the thorax

The unaffected muscles eventually undergo contracture The paralyzed muscles are placed under tension which exerts an injurious effect on them As the result of excessive fatigue and strain weak muscles may become paralyzed long after the active disease has ceased In limbs with muscles paralyzed sufficiently to inhibit their use the bones fail to grow and they undergo decalcification causing an increased excretion of the calcium in the urine

EXAMINATION On physical examination it may be observed that the patient lies quietly in bed and does not wish to be disturbed There are irritability and resentment against being moved or handled The painful sensitivity of the peripheral tissues indicates the presence of spasm a common and useful observation Stiff neck stiff back and shortened hamstrings are familiar examples of symptoms caused by the disease but muscles elsewhere in the body may also be involved by spasm The muscular tenderness occurs in the nonparalytic as well as the paralytic individuals

When the examiner places the hand beneath the patient s head and attempts to flex the neck resistance will almost invariably be encountered as early as clinical symptoms have become manifested An attempt to flex the body will disclose stiffness of the back When the trunk is flexed the knees will rise from the bed because of shortness of the hamstring muscles The stiffness is due to the condition of spasm in the posterior muscles On search the same hypertonicity and shortening may be found although less frequently in other muscles of the body and in all types of cases

The patient may complain of pain in the muscles involved by spasm or pain may be elicited by gently stretching the affected muscles Involvement of the muscles of any part of the respiratory mechanism hampers the breathing Spasm may affect the abdominal muscles

Spasm occurs more frequently in the paralytic than in the nonparalytic patient The value of this observation for the purpose of diagnosis is diminished by the fact that paralysis is not a common early condition and may not occur at all Paralysis occurs at some time within 15 days of onset in two thirds of the cases Neither weakness nor paralysis appears as an initial symptom as a rule and in only a few cases does this condition develop during the first 24 hours

Patients who have bulbar symptoms such as difficulty in swallowing nasal regurgitation facial weakness and difficulty in speech reveal muscular dysfunction on the average slightly earlier than patients in whom the spinal type of paralysis develops

It may be observed that there are twitchings in certain muscles and that in these same segments there is an increase of tendon reflexes accompanied by pain and tenderness In some muscles the reflexes are diminished or lost and weakness and paralysis are present The paralysis may be found on the first second or third day of the illness but it may be delayed until the seventh or eighth day The paralysis may begin in the legs and ascend to involve groups of muscles higher up It may be observed that the paralysis after remaining stationary for several days or weeks has extended once more *In the early stages of the disease* ankle clonus or even plantar extension may be found

In the leg the quadriceps femoris the peroneals the flexors of the foot and the extensors of the toes are more commonly involved than other muscles The iliopsoas and the gluteals are often selected In the upper extremity the deltoid the spinati the flexors of the elbow and the supinators of the forearm are often involved Paralysis of the muscles about the shoulder joint and upper arm is common

The diaphragm is usually affected in association with paralyses of the arms and the muscles of the shoulder girdle In cases of diaphragmatic paralysis breathing is thoracic with the epigastrium retracted during inspiration If the intercostal muscles

tion with poliomyelitis and has some characteristics in common with that disease These viruses differ immunologically from poliomyelitis viruses and do not cause the lesions of poliomyelitis when injected into monkeys The role of the newly discovered viruses in the causation of human disease is still uncertain

Tetanus (*Lockjaw*) In contrast to degenerative or destructive lesions of neuro muscular apparatus involving the muscles as in the dystrophies or the nerves primarily or the cells of the spinal cord as in acute anterior poliomyelitis is the hyperirritability of muscles in tetanus or lockjaw However the very early manifesta tion of both of these diseases is muscular spasm

As a rule *Clostridium tetani* does not multiply within the tissues except in the presence of necrotic material Thus puncture wounds and buried splinters offer favorable condi tions for a focus of this infection The organisms do not reach the nervous system The toxin reaches the nervous system by extending inward along the nerve trunks Once within the central nervous system the toxin diffuses rapidly However according to Abel the toxic conditions caused by tetanus are created by three processes (1) a local lymph borne intoxication of the muscles or motor nerve endings (2) a generalized effect of the same nature due to blood borne toxin and (3) a series of symptoms due to the action of the blood borne toxin on the anterior horn cells of the spinal cord (see Chapter 18)

The *incubation period* of tetanus averages about 7 to 8 days but may be pro longed to several weeks or even months in cases in which antitoxin has been ad ministered One attack does not necessarily bestow immunity

SYMPTOMS The first characteristic symptom of tetanus is tightness and twitch ing of the muscles There is trismus of the muscles of mastication The posterior cervical and abdominal muscles are affected early

In the localized forms of the disease the first symptom is a constant spasm of the muscles in the neighborhood of the wound The spasm extends and frequently involves a whole limb The symptoms may gradually advance for a week or two and the intense muscular spasm then may persist for two months Local tetanus is less serious than the general form

Tetanus infections of the face neck or head may produce a cephalic type of tetanus It begins with spasm of the facial muscles on one or both sides or with involvement of the jaws or pharynx Cephalic tetanus may remain localized or may be followed by a generalized tetanus In a few favorable cases the spasms grow less frequent and severe and the rigidity diminishes in a few days The trismus is often the last sign to disappear Convalescence is slow When the disease is fatal death occurs in 3 or 4 days In some cases of fulminating tetanus death may result from asphyxia within a few hours The temperature often remains normal until toward the end The development of high fever is a bad prognostic sign

EXAMINATION On repeated examinations the tightness of the muscles is ob served to increase. Pressure over the affected muscles and efforts to stretch them cause severe pain As the disease progresses it is observed that the muscular move ments grow more and more limited and finally it is observed that constant and in tense contracture of the muscles has developed so that the muscles have become immobile with spasm The spasm progresses successively to the muscles of the spinal column the abdomen the thorax and the legs and to the arms When all the muscles are involved with spasm the legs are extended the head is retracted and the trunk is arched backward into a position of opisthotonos The arms are flexed and the fingers clenched The angles of the mouth are retracted (risus sardonicus) the eyelids partially closed and the eyebrows elevated By this time the bulbar muscles are affected and dysphagia aphonia and cyanosis are present

Owing to sphincteral involvement there is retention of urine and feces The patient at this stage perspires profusely If contracture of the muscles of respiration or adductor spasm of the larynx occurs fatal asphyxia and death ensue

The generalized tonic spasms of the entire bodily musculature become more frequent and more prolonged The pain is horrifying to the patient and its apparent

The patient who has rheumatic fever may not be able to move the legs because of pain the immobilization however is voluntary There are present pain at the insertion of the tendons or in the joints involved perhaps a cardiac murmur and a good therapeutic response to salicylic acid

In postvaccinal encephalitis there are present encephalitic irritability followed by stupor a history of vaccination and a relatively low *lymphocyte count with an* extremely high protein content in the spinal fluid If the patient is seen at the onset there are signs of an upper but not of a lower motor neuron lesion Later during coma neurologic reactions may be absent

In lymphocytic *choriomeningitis* (*acute benign idiopathic meningitis, acute* aseptic meningitis benign aseptic choriomeningitis epidemic meningitis serosa or serous meningitis) the onset is acute and the progress rapid and it occurs in any age group at any time and in any locality It is accompanied by headache muscular pains and sometimes constipation Neither muscle weaknesses nor neurologic signs are present except perhaps in a positive Kernig sign and a stiff neck Examination of the spinal fluid reveals normal sugar and chloride negative reactions to all tests, a meningitic gold curve a cell count ranging from 20 to more than 1 000 cells per cubic millimeter 90 per cent of which are lymphocytes with little or no protein It is therefore evident that it is impossible to separate this disease from an abortive type of poliomyelitis when it occurs during an epidemic of poliomyelitis

The spinal fluid in equine encephalomyelitis usually is clear to ground glass in appearance and contains from 200 to 2 000 cells per cubic millimeter mostly polymorphonuclear leukocytes the globulin is 4 plus

Tuberculous meningitis in its inception is difficult to differentiate from polio myelitis Time may *be required for the differentiation* A patient who has tuberculous meningitis remains ill for days with but few symptoms and little or no fever and with no paralysis of muscles Specific organisms are present in the spinal fluid

The Coxsackie Viruses Investigation of small epidemics of poliomyelitis led *to the isolation by Dalldorf and Sickles from fecal specimens of 2 children* in the acute phase of an agent that induces paralysis in suckling mice and hamsters This paralysis is associated with destructive lesions of skeletal muscles the central nervous system being unaffected

Howitt reported isolations of the Coxsackie group of viruses from the secretions or tissues of patients from Alabama Colorado Delaware Florida Georgia Louisi ana Oklahoma South Dakota and Tennessee

The Coxsackie virus may be isolated from human feces urine nasopharyngeal or oral washings oral vesicles nasal swabs saliva sputum *serum whole blood* cord and brain The virus has been isolated in sporadic cases and from groups of patients who had variable and ill defined symptoms such as mild fevers polio myelitis like or influenzal syndromes and occasionally encephalitis

Howitt s study revealed Coxsackie virus in the *tissues* and secretions of 10 patients whose disease was fatal in 5 of 10 cases the diagnosis was poliomyelitis Poliomyelitis virus was recovered also from the spinal cord of 1 of these patients and from the feces of 2 patients who had recovered Howitt and Benefield were able to differentiate the Coxsackie virus into serologically *distinct types by the* complement fixation test with filtered muscle antigen The same test can be em ployed for determination of the presence of antibodies in human serums At least two immunologic types of the virus exist The virus has been isolated from the sewage of a number of cities and *from flies collected in widely separated areas*

Findlay has cited evidence to suggest that Coxsackie viruses 1 and 2 are widely distributed in Great Britain and that some epidemics of Bornholm disease (epidemic pleurodynia) in that country are associated with infection by Coxsackie virus 1 or 2 or closely allied viruses It is therefore *apparent that there exists* a common infectious disease caused by the Coxsackie virus group that occurs in close associa

more there may remain a detectable defect in conjugate deviation of the eyes toward the paralyzed side

The upper part of the face is but slightly weakened The facial weakness is more evident on voluntary than on emotional movement At first it may be impos sible to wink the eye of the affected side without closing the opposite eye Voluntary movement is most severely restricted or abolished in the arm Both the arm and the leg reveal the hypertonicity and reflexes characteristic of an upper motor neuron lesion There is weakness of trunk muscles revealed by the inability of the patient to sit up in bed and often to turn in bed There is a tendency to fall toward the para lyzed side when standing is permitted or attempted

The affected arm slowly adopts an attitude of general flexion and adduction The leg develops an attitude of extension and outward rotation The foot is plantar flexed and it so remains The demented hemiplegic patient tends to lie on the con tralateral side which permits flexion of the ipsilateral leg Flexion of both legs ensues because of disuse too of the contralateral leg This attitude of flexed legs becomes fixed unless prevented by braces The addition of braces makes it very difficult to give nursing care

On examination when the hemiplegia has become fully established the findings are those of the upper motor neuron lesion Isolated movements of the fingers and all skilled movements of the hand extension of wrist and fingers extension at the elbow clenching of the fist and flexion at the elbow are affected Elevation and adduction at the shoulder are but slightly impaired Dorsiflexion of toes and foot eversion of the foot flexion at knee and hip plantar flexion of foot and toes and least severely extension at knee and hip and inversion of the foot are reduced or abolished

The movements of the lower part of the face are more severely impaired than those of the upper part Deglutition is unaffected but articulation may be transiently or permanently disordered

The increase in muscular tone is more evident in the musculature of the arms and legs The increased tone or spasticity however is variable in the muscle groups For instance in the upper limbs it is most marked in the flexor muscles of forearm and arm and in the adductors of the shoulder In the lower limbs it is characteristi cally most marked in plantar flexor and extensor muscles Its presence is detected by passive stretching of the muscles and observing an increased resistance An in creased resistance is easily elicited in the knee extensors While the patient is reclin ing in bed request him to rest the thigh upon the examiner s forearm the examiner s other hand grasps the leg just above the ankle and flexes the leg at the knee Slight flexion is normally permitted without marked resistance However as flexion is increased an active resistance develops quickly in the quadriceps muscle and more force is needed to continue the flexion After the knee is flexed to approximately 45 degrees the resistance ceases and the rest of the movement is carried out normally In spastic paraplegia it may be difficult to flex the leg passively In instances of mild hemiplegia only mild muscular hypertonus may be appreciable in the flexors of the fingers and in the plantar flexors of the foot

The loss of cutaneous reflexes is not an invariable feature of upper motor neuron lesions The extensor (Babinski) type of plantar response is present in an upper motor neuron lesion This reflex may be obtained from both cutaneous and deep stimulation Pinching the skin of the foot or leg pressure over bony points around the ankle or a stroking pressure over the shin may elicit the response Thus Oppen heim s Gordon s and Chaddock s reflexes are not more than the extensor plantar response elicited from different parts of its wide receptive field The extensor type of plantar response in those who have paraplegia in-extension may be accompanied by plantar flexion of the foot and great toe of the opposite side the so-called crossed plantar response

intensity defies description for the patient does not lose consciousness The spasm may be so great that fractures of the vertebrae ensue A moderate leukocytosis is present The spinal fluid is normal The pressure is increased

DIAGNOSIS The history of a wound the trismus the opisthotonos, risus sardonicus and abdominal rigidity and the tonic muscular spasm persisting between the paroxysms are diagnostic

There is a mortality rate of less than 50 per cent if patients are properly treated with antitoxins

In treatment for tetanus an antitoxin is used The American unit of antitoxin is defined as 10 times the least amount of serum necessary to save the life of a guinea pig weighing 350 gm for 96 hours against the standard test dose of toxin The standard dose of toxin is 100 M L D of a standard toxin preserved at the National Institute of Health

The Upper Motor (Pyramidal) Neurons and Locomotion It has been indicated in the preliminary definitions that upper motor neurons at least a few of them arise from the Betz cells of the cortex and pass uninterruptedly to the anterior horn cells of the cord It is for the purpose of definition that these neurons have been singled out for in reality the pyramidal system is a twofold communicating system It is an internuncial path interposed between the distance receptors and their central cortical connections and the motor mechanisms of the nervous system Through the pyramidal system pass all the impulses which instigate willed movements of skeletal muscles These movements are activated and designed by the cortical (frontal lobe) afferent patterns of excitation

As the complexities of the nervous system prevail it must be added that in the manifestations of the lesion of the upper motor neurons there are evidences also of overaction in subordinate extrapyramidal neurons This overaction of the extra pyramidal neurons is manifested by spasticity increased tendon reflexes clonus and the extensor type of plantar response These reflex changes have their origin as the result of a loss of influence of the pyramidal neuron The symptoms are often designated release symptoms

Hemiplegia Hemiplegia represents a common acute or chronic manifestation of an upper motor neuron lesion Hemiplegia is caused by a lesion of the brain on the side opposite (contralateral side) the paralyzed muscles of the face arm and leg Usually developing acutely it passes through an acute phase and then subsides leaving the muscles hypertonic or spastic—the spastic phase Initially the onset is sudden and the patient is found unconscious lying with the head and eyes turned away from the side of the lesion Often there is a slight ptosis of the eyelid on the affected side The cheek on the paralyzed half of the face flaps in and out with each breath If the breathing is stertorous the increased amount of sputum is spattered over the face clothing or bedding The entire musculature is flaccid The flaccidity of the affected side of the mouth and the thigh is most noticeable The paralyzed thigh muscles spread out as the patient lies supine and give this part of the limb a broader appearance than the other one The tongue may be thrust out toward the hemiplegic side It is impelled outward by the genioglossus muscle of the normal side The tongue returns to the mouth within an hour or two except in those patients who remain severely paralyzed The base of the tongue often remains higher on the paralyzed side than on the opposite side

For a week or more the temperature of the hemiplegic half of the body may be from 1 5 to 2 F higher than of the opposite half Often acute trophic lesions develop edema of hand and foot with blistering and bedsores The abdominal reflexes are abolished on the paralyzed side Only in the most deeply comatose patients are the tendon jerks abolished The plantar response is extensor on the affected foot The shock usually passes off after a few hours and the patient regains consciousness and on restoration of consciousness release symptoms appear The deviation of the head and eyes away from the side of the hemiplegia passes off but for a week or

be local invasion from osteomyelitis of the vertebrae or from furunculosis with an ensuing septicemia Purulent myelitis and even abscess of the spinal cord may occur in various types of septicemia but these are rare

Tuberculous spondylitis may cause sudden paraplegia although as a rule the onset is slow

Acute spastic paraplegias in association with other types of infection such as those due to the viruses of measles vaccinia varicella or mumps may appear without any other evidence of involvement of the nervous system An acute para plegia may follow a febrile illness of obscure nature or may occur without apparent cause

Paraplegias in-extension due to pyramidal (upper motor neuron) lesions in the cord develop slowly The toe begins to drag and it is frequently stubbed The sole of the toe of the shoe is worn on the inner side This difficulty in locomotion is enhanced by the flexion weakness of the leg and shuffling gait ensues Spasticity develops in the extensor and plantar flexor muscles The plantar response often is extensor in type The legs come to be rigidly extended and abducted Paroxysms of spontaneous clonus or sudden single extension spasms are experienced

Paraplegia in flexion occurs when there is a more complete interruption of conduction of descending impulses in the cord than is present in paraplegia in extension

Involuntary flexor spasms increase and finally the legs come to lie flexed with the knees pressed against the abdominal wall If the legs are forcefully extended they rapidly resume flexion The extensor type of plantar response may be obtained from the middle of the thigh downward by pinching or pricking the skin or deep stroking over bones and muscles The knee jerks and later the ankle jerks may diminish and become difficult if not impossible to obtain

The diagnosis is established by the history and the physical findings Identifica tion of the organism by bacteriologic culture is helpful in treatment

Injuries of the spinal column may involve the spinal cord as a result of hemor rhage or compression by dislocation of intervertebral disks or the vertebrae them selves Often in instances in which the injury of the cord is due to trauma dislocation of the intervertebral disks or fracture of vertebrae may not be demonstrable on roentgenologic examination Such lack of evidence of injury is explained by the fact that dislocations of the vertebrae are reduced spontaneously In rare instances dis location of a vertebra (the first or second cervical) has occurred spontaneously and such an occurrence may cause paraplegia or may cause sudden death

During the course of *multiple sclerosis* especially in association with an optic neuritis from the same cause (neuromyelitis optica) a spastic paraplegia may occur In these cases the paraplegia may improve

The diagnosis depends on the association of acute transverse myelitis optic neuritis and other symptoms of multiple sclerosis

Paraplegias of gradual onset may be due to *extradural sarcomas and gliomas* fibromatous tumors of Recklinghausen s disease syringomyelia tuberculous spondy litis and severe kyphosis of nontuberculous origin

The diagnosis of these conditions depends on expert interpretation of myelo grams made with experienced hands

Extrapyramidal Neurons and Locomotion It has been stated that the extra pyramidal tract consists of short neurons intercommunicating with parts of the pyramidal system (upper motor neurons) and that this system is presided over in some degree by the pyramidal system These short tracts connect with the corpus striatum the substantia nigra corpus subthalamicum red nucleus vestibular and reticular nuclei and long descending tracts the rubrospinal vestibulospinal and reticulospinal

The essential clinical components of the different syndromes of extrapyramidal

When the hemiplegic patient yawns or stretches as during any forceful or sustained movement of the normal limb the affected arm tends to extend at the elbow wrist and fingers, remaining rigidly in this new attitude until the yawn passes off Simultaneously the leg goes into rigid extension with the foot plantar flexed These phenomena are not associated movements but rather are phenomena in the normal maintenance of posture

Due to the fact that the arm and hand have been freed of locomotor functions their movements are controlled from the cortex and are therefore largely acquired or learned movements The degree of loss of movement is permanent The hand rarely regains any movements other than simple flexion of all the fingers simultaneously and simple movements of flexion and extension at the wrist The movements of the face and leg are more primitively spontaneous and natural and thus more recovery will take place in them than in those of the arm and hand The extensor (Babinski) plantar response on the paralyzed side however remains freely elicitable Hemianesthesia with or without a crossed homonymous hemianopsia may accompany the hemiplegia This occurs only when the posterior limb of the internal capsule is extensively destroyed the sensory radiations which lie caudal to the pyramidal fibers and the optic radiations which are placed still further caudally are affected

There is an upper and a lower motor neuron type of paralysis of ocular movements In paralysis of the upper motor neuron there is a loss of co ordinated movements In paralysis of the lower motor neuron there are characteristically present diplopia and squint unless there is complete external ophthalmoplegia The upper motor neuron type of paralysis is a supranuclear paralysis of which there are two varieties one when the voluntary initiation of conjugate movements is lost but *such movements can still be evoked by reflex stimulation the other when such* movements are totally lost and cannot be elicited In total blindness resulting from bilateral destruction of the visual cortex the pupillary reactions may remain intact The lower motor neuron type of ocular paralysis does not occur in hemiplegia

Paraplegia Paraplegia is a paralysis of the lower part of the body and the legs caused by disease or injury of the upper motor neuron in the spinal cord Both motion and sensation are affected Paraplegia may exist in extension or in flexion

Common causes of paraplegia are defective development of the spinal cord infections injury to the spinal cord locomotor ataxia transverse myelitis chronic alcoholism malaria and pernicious anemia

Defective development of the spinal cord or myelodysplasia with spina bifida as has been stated in relation to lower motor neuron lesions usually causes flaccid paralysis and atrophy of the legs Rarely will a defect in the cervical segments of the cord produce spastic paralyzed legs with knee jerks increased Frequently in the presence of spastic paraplegia there is some atrophy of the muscles of the hands and forearms and perhaps loss of pain and temperature sensibility as in syringomyelia

The diagnosis of these congenital lesions of the pyramidal tract with spastic paralysis is made by physical examination and by the finding of an area of defective cutaneous development overlying defective vertebrae demonstrated by roentgenologic examination

In some instances vascular anomalies such as congenital arteriovenous fistulas or venous angiomas may cause sudden spinal paralysis from pressure or rupture Occlusion of the abdominal aorta at the bifurcation may cause anemia of the cord and sometimes paraplegia

Pyogenic infections having produced a pyemia may originate an *infection of the epidural space* causing either a granulomatous process with compression of the cord or an extensive purulent process with a rapidly ascending paralysis which is almost always rapidly fatal The other causes for infectious spastic p[illegible]

are accompanied by great excitement and often delirium These movements fulfill the requirements for an extrapyramidal origin arising in the short extrapyramidal neurons in the cortex

The Cerebellum and Locomotion The cerebellum functions with the pyramidal system in the maintenance of posture The harmonious control of the combination of movement and posture is a function of the cerebellum When there is a loss of function of the cerebellum voluntary movement is grossly disordered and cerebellar ataxia ensues

Cerebellar Ataxia Cerebellar ataxia according to Walshe is a disturbance of postural fixation In acute cerebellar ataxia there are flaccidity and atonicity of the muscles In chronic cerebellar ataxia there is fixation of the parts while actually in movement as well as of the parts which should be maintaining attitude in the rest of the musculature The true expression of a cerebellar ataxia is a defect in tone of the muscles This tonal defect is expressed by abnormalities in the movements of the eyes of the muscles of articulation and of the hands arms and legs

The ocular movements are manifest on conjugate deviation to the side There is a series of regular horizontal jerks nystagmus The eyes can move normally laterally The abnormality is evidenced by *inability to sustain the lateral deviation* The eyes drift slowly toward the midposition During the slow movement of the eyeballs back to midposition they are suddenly jerked back to the deviated position In unilateral cerebellar abscess and auditory nerve tumors the nystagmus is slower and its range wider when the eyes are moved toward the side of the lesion than when they are turned away from the side of the lesion

The movements of the muscles of articulation are so affected as to produce scanning speech In scanning speech each syllable is pronounced deliberately and slowly Movement of the hands reveals an intention tremor irregular in rate and amplitude and excessive in range

The abnormality of the movement of the hands and arms is easily demonstrated by having the patient touch the index finger tip to the nose The hand and arm commence their course normally but soon an irregular course begins with a to and-fro jerky movement which increases in width as the finger approaches its destination The finger finally punches or misses the nose and for an instant continues to wiggle about the end of the nose All movements of the hands are likely to be forceful and to overshoot the mark

Because of irregular swaying of the body the gait is irregular and staggering and the maintenance of stance difficult It seems that the arms and legs are not ready to stop on command Sensations are undisturbed The tendon jerks are exaggerated In unilateral lesions the postural disturbances are limited to the side of the lesion

Patients who are recovering from debilitating illness present a deceptive appearance of cerebellar ataxia which is due chiefly to weakness

CONGENITAL ATAXIA Congenital cerebellar ataxia due to cerebellar aplasia becomes evident in the first two years of life There is ataxia with intention tremor of the arms and unsteady gait of cerebellar type The symptoms remain unchanged throughout life

ACUTE ATAXIA Acute cerebellar ataxia may occur as the only manifestation of encephalomyelitis due to measles varicella and other acute infectious diseases Cerebellar ataxia may occur in association with any widespread lesions in the nervous system Cerebellar abscess is a cause of acute cerebellar ataxia There are usually present of course otitis and mastoiditis as well as fever leukocytosis and intracranial hypertension

Cerebellar Deficits In the syndrome of generalized cerebellar deficit there is progressive unsteadiness of gait Later the co ordination of the legs and arms is impaired Finally static equilibrium is involved and speech becomes dysarthric This syndrome combines the features of the pure cerebellar syndromes and results from a

disease of the motor system are disorders of muscle tone and involuntary move ments Walshe has *described these as three main symptom complexes* (1) the syn drome of paralysis agitans (tremor like rigidity) (2) athetosis and (3) the chorei form syndrome

Paralysis Agitans (Tremor like Rigidity) In paralysis agitans *according to* Walshe there is a uniform muscular hypertonus throughout the entire range of passive or active movement of all muscles except external muscles of the eye On palpation the muscle has an increased tonus which yields only when an appropriate force is applied On forcefully stretching such a muscle as on extension at the elbow *there is a jerky relaxation simulating the lowering of a lever by a cogwheel* thus the term cogwheel rigidity This rigidity slows the speed and the range of movement so that facial expresion lessens and finally disappears giving the patient a masklike rigidity of expression Excessive emotional stimuli are required to break up this rigidity When the patient smiles the smile moves slowly over the face and *once it has spread it remains too long In walking the arms cease to swing and the* legs move slowly and in short steps (loss of associated movements) There is a tremor which is a rhythmic movement In mild extrapyramidal involvement the tremor ceases when the patient is at rest and even *in severe instances of involve* ment it ceases while the patient is asleep Occasionally it may cease while the limb is *in active use* During stresses of emotion or in fatigue the tremor increases The tendon jerks are increased but clonus is absent The plantar response is flexion Occasionally the tremor is entirely absent throughout the course of the illness

The common form of paralysis agitans or Parkinson s disease affects aging individuals as a part of the manifestations of arteriosclerosis Less commonly in its most highly developed form the disorder is a manifestation of chronic encephalitis subsequent to an acute encephalitis a viral disease which occurs sporadically endemically and epidemically (Von Economo s encephalitis) (See Chapter 15)

Often the disease begins with aching in the arms to be followed by tremors of the fingers The tremors may be limited for months to a hand or a foot In due time there is a manifest hypertonia of the muscles more definitely expressed in the flexors than in the extensors As soon as hypertonia is established the head and body become bent forward and there is as well a slight flexion everywhere *in all of the muscles The gait is slow and shuffling and often the patients complain of propulsion* or retropulsion The voice is high pitched weak and monotonous There is difficulty in beginning to speak but once started the words are uttered rapidly

Many and varied are paresthesias and aching pains Disturbances in sweating are common The face is often flushed and expressionless or masklike

The tremor advances until it finally is slowed after the patient is confined to bed Utter helplessness is often present just prior to death

Athetosis The variations in the muscular hypertonus of extrapyramidal [illegible] seem in some instances to be the cause of slow twisting movements and di[illegible] postures of the arms and legs with fingers hyperextended These movements are *aggravated in force and amplitude when movement is performed when the p*[illegible] is hurried or emotionally tense No signs of upper motor neuron lesion may be [illegible] except *in those who have in addition an infantile hemiplegia* The movements of the face tongue and articulation are not affected except in rare forms of bil[illegible] athetosis

The Choreiform Syndrome In contrast to the movements of paralysis ag[illegible] and athetosis choreiform movements are associated with diminished muscular [illegible] These movements *are superimposed upon voluntary movements* They are jerky co ordinate movements of the limbs trunk and facial muscles The involunt movements of rheumatic chorea *are not associated with extrapyramidal disord* and therefore are described elsewhere in this text (Chapter 18) In severe chor such as chorea gravidarum the movements are incessant of great violence a

vidual has good vision and is able to see written words but does not understand them the condition is termed *word blindness* If an individual does not comprehend spoken words there is a *sensory aphasia* If there is only injury to a motor eupraxic center the speech musculature is incapable of forming words though all other movements in which it functions are normally performed the condition is *motor aphasia* When writing movements cannot be made there is *agraphia* A failure to recognize the nature or use of a familiar object such as a spoon is *agnosia* An individual who has neither paralysis ataxia of movement nor sensory loss yet cannot perform the movements appropriate to the use of a familiar object has *motor apraxia*

Sensory aphasia consists in an impaired or lost comprehension of the spoken word expression through speech is disordered It is postulated that the eupraxic center transmits the wrong speech patterns to the motor cortex and thus wrong words are uttered These words are often badly arranged and the patient being word deaf is unaware of the mistakes One affected with word deafness cannot carry out spoken orders

Motor aphasia in its mildest form consists in a loss of memory for the names of familiar objects In more severe degrees of aphasia the patient is reduced to the utterance of a single word such as Yes or perhaps Yes yes (See Diseases of the Larynx Chapter 4)

In aphasic subjects there is a tendency when some other movement is asked for to repeat a movement that already has been carried out to order

Agnosia and *apraxia* are detected by giving the individual familiar objects and thus determining the capacity to recognize their nature and use *Motor apraxia* is detected by the presence of an inability to perform familiar but simple movements

THE SENSORY SYSTEM AND LOCOMOTION

All acts of locomotion are initiated by the sensory nervous system The organs of sense namely the eyes ears nose taste buds touch spots in the skin and the nerve beginnings in the muscles bones joints and viscera are so constructed as to analyze the environmental energy and select certain kinds of disturbances which are then transformed in the receptor and give rise to nervous impulses These nervous impulses on arrival in the appropriate area in the brain originate visual auditory and olfactory sensations taste dermal sensation positional sense of an extremity or pain Aberrations or false interpretations of these sensations by the brain have been considered in association with the complaint Likewise since pain is the commonest subject of complaint the sensory perception of pain was differentiated from the reaction to pain All fibers carrying all of the sensations from the various end-organs in the skin and deeper skeletal tissues arrive at the dorsal root entry zone of the spinal cord Here the modes of sensation are grouped or integrated according to quality Some impulses probably go no higher than segmented levels of the cord The impulses for tactile pressure postural and vibratory sensibility course up the dorsal columns forming the uncrossed sensory paths ending in the dorsal column nuclei of the medulla Then secondary neurons carrying the impulses through the decussation to extend upward traverse the pons and the midbrain and end in the lateral nucleus of the thalamus In the lateral thalamic nucleus all sensations are again integrated and from this nucleus the third and last neuron carries the remaining impulses which were not integrated in the thalamus to the sensory cerebral cortex and to consciousness

The superficial and deep impulses for painful and thermal sensations on reaching the cord enter the gray matter of the dorsal horn to join a secondary neuron which crosses the cord to reach the lateral column of the opposite side where it turns upward Painful and thermal sensations thus result from nervous impulses that follow crossed sensory paths (spinothalamic tract) Tactile sensibility too has

generalized cerebellar damage The chief differential diagnosis is from the spinal ataxias

Cerebellar catalepsy and the rare cases of cerebellar fits tonic in type often with opisthotonos are indicative of disease of the subcortical cerebellar nuclei

In vertigo of cerebellar origin the surrounding objects appear as if rotating toward the side opposite to the lesion However this vertigo may be due to affection of the adjacent vestibular nuclei or their connections Likewise the upward and backward and lateral positions of the head are probably due to vestibular disturbances and not to primary cerebellar disease Implication of the vestibular fibers produces vertigo and spontaneous nystagmus laterally and vertically together with impairment of thermic nystagmus and of other vestibular reactions on the affected side Occipitofrontal headache appears Later a vestibular attitude of the head may develop and there are signs of cerebellar compressions consisting of a reeling gait with a tendency to fall toward the side of the lesion As the tumor enlarges other cranial nerve palsies develop A facial nerve palsy is a sign of great localizing value when present

There may be subjective tingling or neuralgic pains in the distribution of the trigeminal nerve Paralytic changes of the trigeminal nerve are evidenced by decrease or loss of sensation in the trigeminal area and loss of the corneal reflex on the side of the lesion Glossopharyngeal paresis and affection of the upper roots of the vagus produce unilateral palatal palsy on the side of the lesion

The diagnosis of cerebellar disease is established by the presence of a cerebellar ataxia If the ataxia is due to a degenerative disease signs of intracranial hypertension are not present If the ataxia is due to an expanding lesion such as a tuberculoma or a neoplasm then papilledema headache and vomiting are present If the tumor is situated in the posterior fascia of the cranium there are evidences of involvement of the cranial nerves or there is pressure on the brain stem

In the absence of intracranial hypertension a differential diagnosis includes the possibility of the presence of multiple sclerosis cerebellar encephalitis from any cause and also lesions of the contralateral frontal and the contralateral parietal lobes of the hemispheres of the brain

The Frontal Cortex and Locomotion The disturbed functions of the frontal cortex which affect locomotion are termed eupraxic Eupraxia is inexactness of the reproduction of known co ordinated movements It is the frontal cortex that stores the patterns for the skilled movements and movement complexes particularly of speech and the hand that are learned during life The eupraxic centers cause the motor cortex to initiate voluntary motor activities when such are desired or necessary Therefore a loss of function of the frontal cortex with its eupraxic centers causes not a true paralysis of movement but an inability to initiate movements at will All movements however may still be performed spontaneously when attention is not directed to their performance

The Speech Function Speaking and writing physiologically are movements psychologically they are symbols which express thoughts that refer to things

Disorders of articulation conform to the manifestations of upper and lower motor neuron lesions the particular manifestation depending on whether the lesion is situated in the afferent (sensory) path in the motor cortex or in the efferent (motor) path In cortical association areas articulated sounds heard or written words seen and the sensory impressions left by past speech movements are retained Any lesion cutting off the association areas from the projection areas or a lesion within them isolating one part from another will produce a true disorder of speech The disorder may be sensory (receptive) motor (expressive) or a combination of sensory and motor defects

If there is only a sensory defect the hearing of word sounds is normal but one so affected is unable to understand what is heard and has *word deafness* If an indi

The element of light contact or touch is tested by a camel s hair brush or a wisp of cotton touched lightly to the skin the patient answering immediately on feeling the touch Symmetric parts should be compared The power of localization is deter mined by touching the skin with the fingertip and asking the patient to close the eyes and put a finger on the same point Any error greater than 2 inches is considered abnormal Localization on the unaffected side should be tested

For testing the temperature sense test tubes containing hot and cold water may be used Touch the surface first with one then with the other asking the patient in each case to state whether there is a feeling of hot or cold

A convenient way to test the weight sense is to use two coins differing in weight but preferably of nearly the same size Alternately and synchronously lay them on the extremity or part to be tested and request the patient to indicate which is heavier This sense is most acute on the brow temples forearm dorsal surface of hand and abdomen

The sense of position is tested while the patient s eyes are strictly closed or blind folded Taking hold of one of the extremities flex extend and move it in a variety of directions asking the patient to imitate these motions with the corresponding limb of the opposite side or place one limb in a certain posture and have him describe the position in which it remains

The power of co-ordination depends on the muscular sense by which is ascer tained the amount of strength to be employed and on the articular and tendinous sense which informs the sensorium of the position of the various limbs and parts of the body for instance the regular and smooth co operation of individual muscles or muscle groups which are requisite to accomplish a definite action or movement If these senses are dulled or abolished the condition of *ataxia* exists However in ataxia other factors are concerned such as sight and touch

Ataxia is a disturbance of co ordination of muscular movements and is usually of three types cortical spinal and cerebellar In *cortical ataxia* the conduction fibers between different centers are usually involved as in the speech defects of general paresis In *spinal ataxia* the conducting fibers of deep sensibility are involved In *cerebellar ataxia* the sensory conduction fibers are involved especially those of the vestibular apparatus In ataxias eye control and disturbance of the sense of equi librium and of vestibular control are the most important accompanying factors

Ataxia manifests itself in certain disturbances of station and gait or other vol untary movements These disorders of co-ordination may be searched for as follows Ataxia of the upper extremities is tested by having the patient close the eyes and touch first with one index finger then with the other the tip of the nose the lobe of the ear the center of the closed eye or the end of an indicated finger of the opposite hand or with the eyes open to thread a needle button the coat or write If these attempts are successful the co ordination is good

Ataxia of the lower extremities may be demonstrated by various methods Have the patient stand eyes closed with the heels and toes together In a perfectly healthy person the swaying of the head will be very slight If ataxia is present the swaying will be noticeable and the patient if not prevented may even fall This symptom is called static ataxia or the *Romberg sign* Ask the patient further to walk along a straight line Inability to follow the line signifies motor ataxia In instances of inability to walk ask the patient eyes open to imitate with a foot the movements made by the examiner s hand for instance writing in the air circles or with eyes closed to touch with the heel of one foot the knee of the opposite side (knee heel test)

In all tests for ataxia which are made with the eyes closed the patient should first rehearse once or twice the required movement with eyes open

In cerebellar and some cortical (frontal) ataxias the patient walks in a reeling drunken manner with short steps and feet wide apart With eyes closed the patient

a crossed sensory path (ventral spinothalamic tract) as well as traveling upward in uncrossed tracts

All sensibility is divided into superficial and deep by Walshe who recognizes four primary modes of superficial or cutaneous sensibility, namely light touch cold warmth and pain Likewise deep sensibility is divided into four modes namely, painless painful pressure postural sensibility and vibratory sense

Each mode of cutaneous sensibility begins in the small groups of special end organs and is conducted by its own conducting fibers These special sense organs are at least doubly innervated by the dorsal root fibers so that when sense organs are sufficiently separated in space two sensory stimuli can be felt and distinctly recognized (Walshe)

EXAMINATION The superficial and the deep sensibilities may be tested by various means The *superficial sensibilities* are conveniently tested as follows (1) light touch tested by cotton wool (2) distinction of small differences between the points of a compass (3) localization the patient indicates spot touched by examiner (4) recognition of small variations (2 to 5 degrees) in the temperature of objects (a test tube of warm water) (5) pain sense tested with a needle

Deep sensibilities usually tested are those of muscular articular and tendinous senses as follows (1) position tested by imitating with the sound limb the position of the affected limb by pointing with the sound limb (2) passive movement appreciation of movement recognition of the direction of movements (3) active movements imitation of movement by the sound limb (4) weight sense (a) with hand supported recognition of differences in weight (b) with hand unsupported comparisons of weight placed in each hand (5) sensibility of bone tested by a tuning fork of low pitch over bony areas (6) joint sensibility tested by flexing and extending joints (7) temperature sense tested by hot and cold test tubes

These sensory functions are examined in order to determine whether they are abnormally active absent or perverted In testing the cutaneous and pain senses the patient s eyes should be kept strictly closed or blindfolded and instructions given that the patient make prompt response at the instant the artificial stimulus is perceived using always the same word Yes or No but otherwise keeping silent Promptness of response is important for slowness in response may indicate a delay in conduction The time elapsing between stimulus and response is under normal circumstances one tenth of a second in disease it may be 10 seconds In testing tactile temperature and pain sensibility the areas in which disturbances are found should be outlined In some instances it is necessary to examine not only the cutaneous surface but also the mucous membrane of the nasal and oral cavities and the external genitalia The following methods are useful

In order to determine the sensibility to small differences between points on the skin ordinary compasses with their tips guarded by bits of adhesive plaster a hairpin or the heads of two ordinary pins may be used The patient s eyes being closed the points are placed on the skin sufficiently wide apart to be recognized as a double contact and gradually brought together until it appears to the subject that he is being touched with a single point only Note also whether one point is felt as two or more and whether a touch on one side of a limb or of the body is felt respectively on the other side of the limb or on the opposite side of the body The sensibility varies within wide limits These limits are learned by the examiner through experience

The tip of the tongue is most sensitive it can differentiate between points 1 mm (about 1/25 inch) apart The finger tips are the next most sensitive areas In the following order the distance of point differentiation decreases lips dorsal surfaces of fingers tip of nose forearm tips of toes cheeks eyelids temples backs of the hands neck leg dorsa of feet back, arm and thigh Over the thigh the patient does well to differentiate points 3 to 4 inches apart

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in walking may reel first to one side or the other, or fall forward or backward In the frontal cortical ataxias which closely resemble cerebellar ataxias the symptom known as adiadochokinesia (inability to make certain rapid fine movements of the hands such as five finger exercises or rapid wiggling of the toes) is usually absent In cerebellar ataxias it is more often present

Anesthesia according to a strict definition indicates a loss of light touch sensibility but the term is often employed to denote a loss of any form of sensation The distribution of tactile anesthesia is somewhat varied It may be either psychic or due to some organic lesion In general the most important diseases or conditions which present anesthesia as a symptom are cerebral lesions causing hemiplegia hysteria and traumatic neuroses disease of the posterior column roots or horns of the spinal cord especially locomotor ataxia pressure on the cord (tumor fracture) neuritis (a frequent cause) and leprosy

Hemianesthesia is a loss of sensibility confined to one lateral half of the body When found it is in the majority of cases a symptom of hysteria It usually affects the left side and terminates very exactly at the middle line of the body Not infrequently there is anesthesia of the senses of hearing taste smell and sight on the same side The next commonest cause is a lesion of the posterior third of the posterior limb of the internal capsule at which point the sensory fibers pass upward Under such circumstances it is usually incomplete and is conjoined with hemiplegia of the same side Hemianesthesia and hemiplegia on the same side plus oculomotor paralysis of the opposite side (crossed ocular paralysis) are indicative of a lesion in the crus on the same side as the ocular paralysis Anesthesia of one side of the body with anesthesia of the opposite side of the face (crossed facial anesthesia) may be due to a lesion of the upper portion of the pons Very rarely hemianesthesia is indicative of a lesion in the optic thalamus or a large cortical or subcortical lesion of the parietal temporal or occipital lobes or is associated with multiple sclerosis An incomplete hemianesthesia of one side with partial hemiplegia of the opposite side may occur as a result of a unilateral lesion of the spinal cord Spinal cord lesions cause irregularly distributed disturbances of light touch sensibility especially important in the early diagnosis of spinal cord tumors

Bilateral anesthesia sometimes amounting to a double hemianesthesia but generally confined to the lower extremities and the lower part of the trunk is rarely if ever due to a lesion of the brain It is an infrequent symptom in hysteria Anesthesia of the lower extremities if in conjunction with varying degrees of paralysis is usually due to some lesion affecting the spinal cord such as an injury or compression of the cord by dislocation fracture or caries of the vertebrae spinal meningitis hemorrhage into the cord or meninges, tumor myelitis or locomotor ataxia

Anesthesia of irregular distribution is due as a rule to either hysteria or neuritis The loss of sensibility may involve one arm or one leg or one side of the face alone or may occur in multiple and scattered patches in any portion of the body Anesthesia of a single extremity rarely originates from a cerebral lesion but when arising from this cause the anesthesia is most marked at the terminal portion of the limb and gradually lessens as the trunk is approached if caused by a spinal lesion its proximal border will present a sharp line of demarcation In order to determine whether they are coterminous with the area of distribution of any of the peripheral nerves the shape and location of isolated areas of anesthesia should be compared with a chart to find if they correspond to definite spinal segments Single or multiple circumscribed patches of anesthesia are usually due to hysteria neuritis or tabes dorsalis

An excessive sensibility to tactile and other impressions attends a considerable number of disease conditions and is termed *hyperesthesia* Areas of hyperesthesia are of frequent occurrence in hysteria and constitute the so-called hysterogenic zones for instance tender points on the thorax the lower part of the abdomen and the back pressure on which will excite and if continued may stop a hysterical parox

ysm In neurasthenia there is a frequent hyperesthesia of localized points along the spinal column and on the scalp and thorax The scalp is often tender following headaches (especially of migrainous type) and facial or occipital neuralgias The area supplied by any neuralgic nerve may be temporarily hyperesthetic The para lyzed side in a hemiplegia may be slightly oversensitive and in some cases of brain tumor there is a marked and extensive hyperesthesia In unilateral lesions of the spinal cord there is a narrow zone of hyperesthesia above the level of anesthesia on the side of the lesion

Disturbances of the temperature sense consist in a loss of the cold sense or the heat sense or both or a condition of reversal of sensation cold being called hot and vice versa Such alterations from the normal are especially characteristic of syringomyelia and to a less extent of locomotor ataxia and lesions of the medulla

Disturbances of the pain sense comprise *hyperalgesia* an excessive sensibility to painful stimuli and *analgesia* a loss of sensibility to pain Analgesia is found par ticularly in syringomyelia and hysteria

Loss of the muscular articular and tendinous senses constitutes ordinary versus cerebellar ataxia It indicates an interruption of the sensory conduction tracts for such impressions It is seen especially in lesions involving the incoming sensory fibers as in locomotor ataxia and Friedreich s disease Disease in the ascending tracts myelitic disease compression and occasionally lesions at the higher levels of the medulla and pons may cause it Diffuse lesions as in multiple sclerosis and paresis may also be responsible for it

Of these separate senses loss of weight sense (muscular anesthesia) may be found alone in hysteria and cortical lesions impairment of posture sense (articular and tendinous anesthesia) may be present in tabes and neuritis before ataxia is found static ataxia involves muscular and articular sensation and motor ataxia concerns articular and tendinous sensations

Astasia abasia inability to stand and to walk although the legs are otherwise under control is generally a symptom of hysteria rather than a separate disease and most commonly succeeds an emotional storm or an injury It has been observed also in tabes multiple sclerosis spastic paraplegia multiple neuritis and exoph thalmic goiter It has been known to follow infectious diseases carbon monoxide poisoning overexertion in walking and painful affections of the leg There is no paralysis and while the patient is in bed the movements of the feet and legs show no in co ordination but on attempting to stand or walk the legs give way Women and young adults are commonly affected

Irregularities of sensation such as allochiria which is a transference of sensation so that a touch on one side of the body is felt on the opposite side may be present in hysteria locomotor ataxia disseminated sclerosis and myelitis Polyesthesia (a touch with one point felt as two) is of the same significance Delayed conduction (tactile or pain) so that the response may require 10 seconds instead of one tenth second may be found especially of pain in locomotor ataxia and various peripheral paralyses Of similar moderate suggestiveness are after sensation an increasing pain lasting for some minutes after a pin prick and double sensibility to touch and pain in which the tactile impression is first perceived the painful impression coming to consciousness after a varying time

Stereognosis is the power of recognizing the form or shape of objects when handled *Astereognosis* is the loss of this power and is found in spinal or peripheral lesions attended by complete tactile anesthesia or loss of deep sensibility In cerebral disease it is significant of a lesion (tumor hemorrhage) in the parietal lobe the center for deep sensibility and perception of form

In *paresthesia* the sensations of tingling and numbness may result from lesions of any part of the sensory pathway

In general lesions of the peripheral nerves commonly affect all modes of sensi

bility Localized lesions of the spinal cord may impair some modes while leaving others intact Thalamic lesions may involve all modes though not always equally Cortical lesions tend to involve the spatial and discriminatory elements of sensibility

Spontaneous pain as a result of central lesions characteristically occurs in con nection with thalamic lesions Severe and persisting pain may arise from relatively slight injuries when irritation is set up at the site of injury Pain as the result of peripheral nerve injury may be of an intense burning quality and may be associated with dryness redness and extreme hyperesthesia of the skin innervated by the af fected nerve (causalgia)

The resulting disability from sensory loss varies and is difficult to evaluate The disability and disorder of movement ensuing on defects in postural sensibility are known as *sensory ataxia* There is an unawareness of the position of the lower limbs and of the range and direction of movements The benefits of vision in control of these disorders are remarkable for vision gives direction and to some extent control of movement When the eyes are closed or when the patient is in the dark ataxia develops and the patient begins to sway and stagger a phenomenon familiarly known as Romberg s sign In the upper limbs the subject has difficulty in distinguishing by manipulation the size and shape of objects held in the hand and this is termed astereognosis The finger nose test determines the presence or absence of postural sense in the upper limbs Have the patient endeavor with eyes closed to place the index fingertip on the nose The arm deviates from its normal path moves around in an uncertain way and may fail to reach its objective

In general with cerebral disease incontinence of either urine or feces is the result of disturbances of consciousness The mental apathy associated with frontal lobe lesions often is accompanied by the passage of urine and feces These acts should be termed mental incontinence rather than sphincteral incontinence

In the semicomatose patient a distended bladder often leads to restlessness and indications of distress and therefore the possibility of its presence is manifest In acute transverse lesions causing total paraplegia there is retention of urine followed by overflow and finally by automatic micturition The periodic sphincteral relaxation allows the escape of some of the vesical contents but not of all There is always residual urine In slowly progressive compression of the spinal cord the initial sphincteral disorder is followed by automatic micturition Acute retention of urine may suddenly develop in one of these bladders The paralyzed bladder often be comes infected and chronic infection leads to shrinkage of the organ and to almost continual dribbling of urine and thus the condition grows worse In total paraplegia from transverse lesions of the cord periodic fecal incontinence occurs

THE NONSENSORY SYSTEM AND LOCOMOTION

The pathways of the nonsensory stimuli seem to be the two spinocerebellar tracts of the cord These impulses arise in muscles joints bones and the labyrinths It is believed that these stimuli are integrated by the gray matter which includes the red nucleus and the cerebellum It is therefore conceivable that the cerebellum is the organ the functions of which serve the cerebral motor cortex to adjust movements and attitudes for har monious co ordination of motor actions without cortical directions

DEEP SENSIBILITY

Mechanics of Visceral Pain White states that observations on the pathways of visceral pain in man have revealed that there are sensory nerve endings in the viscera particularly in the heart and intestine The afferent axons similar to those found in cutaneous sensory nerves run in the autonomic nerves from the thoracic and abdom inal organs to the posterior roots of the spinal cord These axons are anatomically and physiologically different from sympathetic and parasympathetic fibers which they accompany in the autonomic nerve trunks These anatomic differences have

been correlated with knowledge gained from experimental stimulation of human viscera the results of resection or chemical blocking of the various autonomic ganglia and trunks and interruption of posterior spinal roots

There are pain sensitive fibers in the vagus and pelvic parasympathetic nerves and in the thoracolumbar sympathetic ganglia and splanchnic nerves

In malignant disease of the viscera intractable pain is often due to direct extension of the growth into the spinal nerves (see Autonomic Nervous System Ch 23)

Tenderness which is a painful sensation produced by pressure is slightly different from *hyperalgesia* Hyperalgesia may be present without causing tenderness In some conditions of severe hyperalgesic pain especially the colics the patients find application of pressure even a strong firm pressure most gratifying Sometimes there is dissociation between the tenderness of subjective pain and the pain itself the tenderness being present over the site of the lesion while the subjective pain may be limited to that region or may be referred to a distant region

There are three painful reactions to pressure (1) The pain is increased by pressure of any kind and the lightest touch causes severe distress (2) The pain is increased by deep pressure only this generally indicates some deep inflammatory lesion (3) The pain is increased by superficial pressure only This is usually a manifestation of a psychoneurosis

Rigidity of the underlying muscles is as a rule associated with tenderness and is the only good confirmative sign that pain is present Points which aid in differentiating the malingerer from the actual sufferer are the characteristic changes in respiration and pulse both in rate and rhythm that occur when pain is produced Often there are varying amounts of perspiration especially of the palms of the hands

THE REFLEXES

The reflexes to be tested depend on the prevailing complaints and manifestations Generally and routinely some of the cutaneous or superficial reflexes the tendinous or deep reflexes and in certain instances the organic or excitoreflex action are tested

All reflexes presuppose the traveling of a stimulus from the periphery along the afferent nerve to the motor cells in the cord or medulla The motor cells transform the received stimulus into an impulse which is reflected to the periphery along an efferent (motor) nerve to certain muscles which in consequence contract involuntarily This reflex action ordinarily occupies from one twelfth to one tenth of a second The superficial or cutaneous reflexes are elicited by irritating the skin or mucous membrane thus causing contraction of the muscles near the irritated part the deep or tendon reflexes are produced usually by striking the tendon but also by sharp percussion of the muscle or the periosteum near the tendon The organic or visceral reflexes involve a definite and co ordinate response to special stimuli as for instance defecation

The Superficial (Cutaneous) Reflexes The cutaneous reflexes are usually tested by sharply stroking the skin with a pointed object or by scratching tickling pinching and pricking When accurately tested the cutaneous reflexes are diagnostically important Table 6 1 gives a list of these superficial reflexes

The presence of a cutaneous reflex is of value because it demonstrates that the reflex arc (sensory nerve spinal segment motor nerve) on which it depends is normal

Of all the superficial reflexes the plantar reflex is of greatest diagnostic significance In order to perform the test the patient must be recumbent and the foot comfortably warm while the limb should be partially flexed at hip and knee and at the same time rotated outward The sole should then be gently stroked from the heel toward the toe (Fig 6 67)

In the normal person in all periods of life after locomotion has been acquired

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The Deep or Tendon Reflexes Tendon reflexes are the fundamental response of muscle tone A tap on the tendon produces a momentary stretch which in turn stimulates the sensory organs in the fleshy part of the muscle

A tendon reflex is abolished by an interruption of its reflex arc either on the afferent or on the efferent side and in total division of the spinal cord The reflex arc may also be interrupted where the afferent and efferent links of the arc meet at the synapse within the spinal cord for example as in an injury a compressing tumor or syringomyelia The segmental localization of the reflex arc is made use of in determining the level of a lesion in the cord

The tendon reflexes are obtained by delivering a quick blow to the tendon with the middle finger of the hand or preferably with a reflex hammer

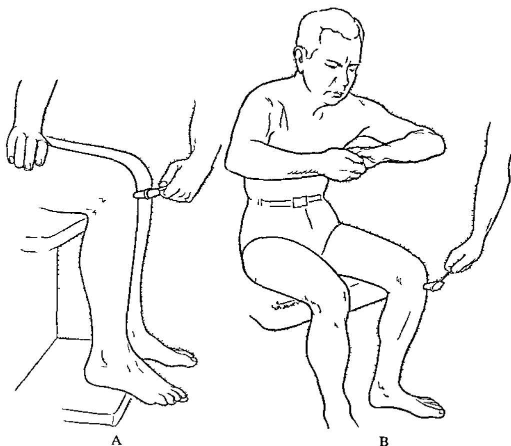


Fig 6 68 A knee jerk Segmental level L_1 L L_3 L_4 B re-enforcement to enhance the reaction

In testing *knee jerk* the leg should be at right angles to the thigh (Fig 6 68A) The knees may be crossed or the examiner may support the leg by placing his hand in the crook of the knee or push his hand from the outside far enough under to rest it upon the opposite knee of the patient thus letting the leg swing upon his fore arm or if the patient cannot sit up the legs should be slightly flexed and supported The patient should be directed or diverted so that the leg is relaxed A sharp blow (with edge of hand finger hammer) should be struck upon the tendon just below the patella Under normal conditions there follows an abrupt jerk of the leg and

Table 6 1 The Superficial Reflexes

Reflex	Method of Eliciting	Response	Segmental Level
Conjunctival	Touch cornea (cotton)	Eyelid closes	Pons
Pharyngeal	Touch wall of pharynx	Pharynx constricts	Medulla
Palatal	Touch soft palate	Palate elevates	Medulla
Scapular	Stroke interscapular region	Scapular muscles contract	C_5 to T_1
Epigastric	Stroke downward from nipple	Epigastrium dimples	T to T_9
Abdominal	Stroke down from costal margin	Abdominal muscles contract	T_{11} to L
Cremasteric	Stroke inner side of thigh	Testicle is pulled up	L_1 to L
Gluteal	Stroke skin over buttocks	Gluteal muscles contract	L_4 to L
Plantar	Stroke sole of foot	Great toe plantar flexed	L_3 to S
Bulbocavernosus	Pinch dorsum of glans penis	Compressor urethrae contracts	S_3 to S_4
Superficial anal	Prick skin of perineum	External sphincter contracts	S and conus

the great toe will be drawn into plantar flexion This reflex depends on continuity between the pyramidal tracts and the spinal reflex Should the pyramidal tract be interrupted by any cause whatever the reflex phenomenon is immediately changed so that instead of plantar flexion there is plantar extension This plantar extension is known as Babinski s sign In infants who have not yet acquired the skilled act of walking Babinski s reflex is normally present The reflexes of Oppenheim Chaddock and Gordon are but variations of or bear the same significance as the plantar response The segmental level of the plantar reflex is L_3 to S



Fig 6 67 Method of obtaining plantar response Segmental level L_3 to S

The palatal reflex may be lost in hysteria and bulbar paralysis The superficial reflexes in general may be absent on the affected side in the early stages of hemiplegia

The conjunctival reflex is elicited by touching the cornea with a soft material such as a wisp of cotton The eyelid closes The reflex has its segmental level in the pons

When the wall of the pharynx is touched immediately the pharynx constricts This reflex has its segmental level in the medulla The soft palate reflex is likewise in the medulla

The scapular reflex is elicited by stroking the intrascapular region The muscles are seen to contract The circuit is to C_5 C_6 C C_8 T_1

The epigastric reflex is obtained by stroking downward from the nipple There is a quick visible response of the muscles The arc is T_7 to T_9

On stroking downward from the costal margin the abdominal muscles contract The abdominal reflex has its segmental level in T_{11} to L

The cremasteric reflex is elicited by stroking the inner side of the thigh just below the level of the pendulous scrotum This reflex has its segmental level in L_1 to L

The superficial anal reflex is obtained by pricking the skin of the perineum its response is contracture of the anal sphincter Its level is S_5 and below

Ankle clonus is a peculiar rhythmic contraction of the calf muscles Partly extend the leg it is essential that the knee be slightly bent Let the heel or the leg rest in the examiner s right hand while his left hand seizes the front part of the foot (Fig 6-70) and abruptly pushes up or dorsiflexes it upon the leg The calf muscles contract forcing the foot downward against the examiner s hand and if the latter continues to press steadily on the sole of the foot a rhythmic clonic movement of the foot will begin and continue In some cases a very small amount of flexion *and pressure will produce it True* ankle clonus must be distinguished from the few clonic movements rapidly ceasing which may be observed as a result of similar manipulations in cases of neurasthenia and hysteria

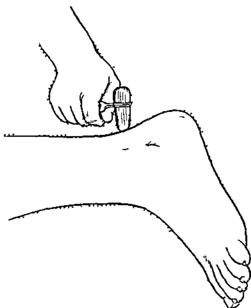


Fig 6 69 Ankle jerk Segmental level S_1 S

Paradoxical contraction is some times observed while testing for ankle clonus in a leg the muscles of which are extremely spastic It consists of a tonic contraction of the anterior tibial muscles produced by the abrupt dorsi flexion of the foot The segmental level of the ankle clonus is S_1 to S

Ankle clonus occurs in those conditions initiating hyperactive knee jerks and ankle jerks Often in a patient who has ankle clonus a similar clonus may be pro duced by sharply pushing the patella toward the patellar clonus

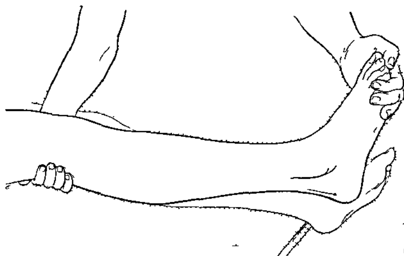


Fig 6 70 Ankle clonus The reflex is obtained while the patient is seated or lying with the knee slightly flexed

There are a few other tendon reflexes which may be used The *jaw reflex* is ob tained by tapping the lower jaw directly with the hammer or the blow may be cushioned as in Figure 6 71 The segmental level of the reflex *is the pons* The jaw reflex is often absent in health It is not a significant reflex

foot If it cannot be obtained, employ re enforcement for instance request the patient to clench his fists strongly or hooking his fingers together to pull one hand against the other using all his strength (Fig 6 68B) Either by lessening cerebral inhibition or by causing an increase of the general muscular tension re enforcement may elicit an apparently absent reflex In some cases of exaggerated knee jerk it is possible to obtain a knee clonus equivalent to ankle clonus by fully extending the leg seizing the patella quickly pushing it downward and continuing the pressure As a result the quadriceps may contract rhythmically for a considerable period

The segmental level of this reflex is L_1 to L_4

Absence of the knee jerk is caused by a lesion affecting any part of the reflex arc Like the other reflexes presence of the knee jerk implies a healthy condition of the afferent (sensory) nerve the posterior roots the intraspinal paths and anterior horn of the cord the efferent (motor) nerve and the muscle itself If the function of any portion or element of this circuit is in abeyance the reflex is modified Consequently loss of the knee jerk is a symptom of disease affecting either the motor or sensory fibers or both Knee jerks are usually absent in neuritis disease of the posterior roots and columns—locomotor ataxia and Friedreich s disease diseases of the anterior horns—anterior poliomyelitis (acute or chronic) and Landry s paralysis and transverse myelitis if affecting the second and third lumbar segments in which the reflex is localized Knee jerks are absent in apoplexy immediately after the shock in epilepsy immediately after the convulsion in injuries to the cord immediately after the accident and in spinal meningitis They are frequently lacking in the toxemias of diphtheria diabetes mellitus (sometimes also insipidus) and chorea

Exaggeration of the knee jerk shows that the reflex arc is intact but that either the normal restraining influence of the upper (cerebral) motor neurons is destroyed by lesions affecting the cells or their fibers which run down in the lateral pyramidal columns are destroyed or that the irritability of the spinal cord as a reflex center has been increased Consequently taking the intracranial causes first the patellar reflex is exaggerated in the hemiplegia of apoplexy on the affected side shortly but not immediately after the attack in the cerebral paralysis of children in hereditary cerebellar ataxia and in general paralysis of the insane The lesions affecting the function of the pyramids are multiple lateral and amyotrophic lateral scleroses Transverse myelitis injuries to the spinal cord (after the immediate effects of the traumatism have passed) pressure on the spinal cord and unilateral lesion of the spinal cord if situated above the level of the second and third lumbar segments will by cutting off the inhibiting cerebral impulses cause exaggeration of the knee and ankle jerks If the lesion is unilateral the exaggerated reflex will be on the same (paralyzed) side The patellar reflex may also be exaggerated as a symptom of hysteria neurasthenia rheumatoid arthritis tetanus and strychnine poisoning

The *ankle jerk* is usually present in health ankle clonus is usually abnormal Absence of the ankle jerk has the same although less important significance as absent knee jerk exaggerated ankle jerk and the presence of the ankle clonus *are* of equivalent value to exaggerated knee jerk the clonus being found *especially in* lateral amyotrophic and disseminated sclerosis A brief abortive or false clonus has been mentioned as occurring in neurasthenia and hysteria

In testing *ankle jerk* extend the patient s leg and hold it up by grasping the *foot* at the same time bending the foot upward so as to stretch the tendo achillis

A better position is obtained by having the patient kneel on a pad or a pillow on a low stool or on the steps of the examining table When in readiness *strike the* tendon sharply and observe the resulting extension of the foot *and the contraction* of the muscles of the calf of the leg (Fig 6 69) The segmental level of this reflex is S_1 to S

response of the tendon reflexes in a limb or between one side and the other is often of diagnostic import when evaluated by an experienced examiner

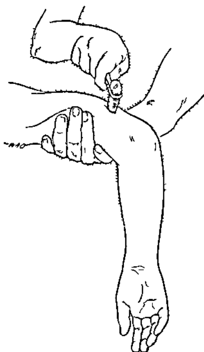


Fig 6 73 Triceps reflex Segmental level C_5 C_7 C

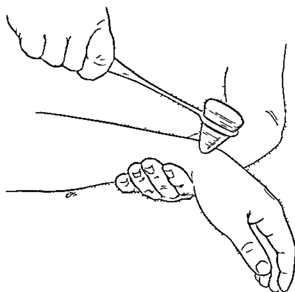


Fig 6 74 Supinator longus reflex Segmental level C C_6

Muscle Tone and the Reflexes Besides the many disturbances of the nervous system and in the muscles themselves muscle tone may or may not be changed If it is increased it is termed hypertonus and if it is decreased there is a hypotonus or an atonia

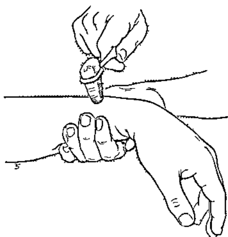


Fig 6 75 Carpometacarpal reflexes Segmental level C_6 C T_1

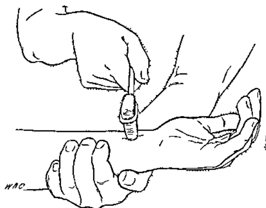


Fig 6 76 Flexor tendon reflex Segmental level C_6 C C_8

A slight but general increase of muscle tone attends preoccupation or emotional states This fact is made use of in the procedure of re enforcement when there is difficulty in eliciting the tendon jerks

The *scapulohumeral reflex* is obtained by tapping the inferior angle of the scapula The reflex level is C_5 and C_6 The supinator longus tendon at the wrist when tapped by a reflex hammer causes a quick but perceptible contraction of the fingers The segmental level is C and C_6

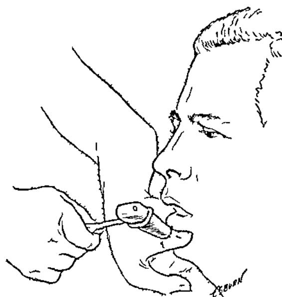


Fig 6 71 Jaw reflex Segmental level the pons

The deep reflexes of the *upper extremities* are of much less diagnostic value than those of the lower extremities because they are frequently absent in health The diagnostic significance of these jerks when present allowing for the difference in localization is the same as that of the knee jerk

The biceps reflex is obtained by tapping the biceps tendon as illustrated in Figure 6 72 The reflex level is C and C_6 The triceps reflex is obtained by tapping the triceps tendon (Fig 6 73) Its segmental level is C_5 C_6 C

The supinator longus tendon reflex may be obtained by tapping the tendon at the wrist (Fig 6 74) The segmental level of the reflex is C C_6

The carpometacarpal reflex is obtained by tapping the back of the wrist The fingers are extended The segmental level of the reflex is C_6 to T_1 (Fig 6 75)

When the flexor tendons at the wrist are hit with a reflex hammer the fingers are observed to be transiently flexed The level of this reflex is C_6 C C_8 (Fig 6 76)

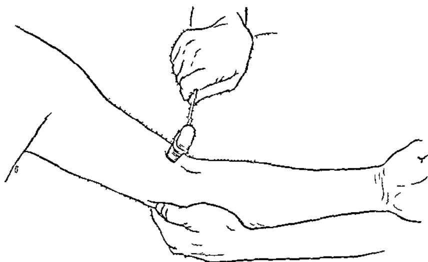


Fig 6 72 Biceps reflex Segmental level C C_6

The *abolition of a tendon jerk* may be associated with the appearance of an alternative response This phenomenon is called inversion of the reflex Inversion may be observed in injuries of the cervical portion of the spinal cord associated with damage to the fifth and sixth cervical vertebrae

What constitutes a normal degree of briskness in a tendon jerk is judged by the experience of the examiner The differences between the rapidity or sluggishness of

lesions commonly are found on the thorax abdomen neck shoulders thighs buttocks and face The lesions are unilateral and rarely involve more than three dermatomes However extensive eruptions do occur Within 2 to 3 days after the zoster a generalized cutaneous eruption resembling varicella may appear Localized paralysis and atrophy in the muscles of the thorax or abdomen corresponding to the distribution of the eruption may develop These palsies are permanent in most instances Symptoms of irritation of the parietal peritoneum or pleura or even of the urinary bladder may accompany zoster Hiccough may occur in association with thoracic zoster

Frequently some of the cranial nerves are involved by zoster Affection and paralysis of the third fourth and sixth nerves are rare The gasserian ganglion and the first branch of the fifth nerve are frequently affected The eruption affects the upper eyelid the conjunctiva and cornea and is accompanied by severe pain The keratitis of zoster may leave permanent scarring of the cornea As the result of anesthesia a neuroparalytic keratitis may ensue Optic neuritis and loss of vision have been recorded

A facial palsy and zoster of the external auditory canal and loss of taste may be associated The eruption of zoster may occur over the anterior two thirds of the tongue and the hard palate or the anterior pillars of the pharynx

DIAGNOSIS The eruption of herpes zoster is characteristic The difficulty and the important diagnostic consideration are the differentiation of primary from secondary forms of the disease The presence of any signs or symptoms which cannot be reconciled with the symptoms of the primary disease arouses a suspicion of some other nervous disease A zoster may be the first intimation of metastatic malignant disease in or along the spinal column or the sequence of irradiation therapy

The prognosis of primary zoster is good Herpes zoster of the gasserian ganglion may cause permanent damage to the cornea and in some cases the facial palsy associated with geniculate herpes may persist The syndrome of crocodile tears may follow herpes of the ear As a sequela of facial nerve zoster there may be profuse lacrimation from the homolateral eye whenever food is taken

Tabes Dorsalis (Locomotor Ataxia) Tabes dorsalis a manifestation of neurosyphilis is commoner in men than in women The disease is manifested from 5 to 20 years after the primary infection by *Treponema pallidum* (see Treponema in Chapter 17) Tabes dorsalis may result from congenital syphilis and may occur in childhood adolescence or early adult life

Regularly there is a low grade inflammation of the posterior spinal ganglia of the roots between the ganglia the spinal cord and the meninges A selective degeneration of the neurons of the posterior columns of the cord is characteristic There may be degeneration of the optic nerve and the nerves supplying the ocular muscles The trigeminal and the vagus nerves occasionally are affected The nerve pathway which has to do with the pupillary reflex is almost always affected early producing the Argyll Robertson pupil

SYMPTOMS As seen in the private practice of medicine patients who have tabes dorsalis usually complain of symptoms which are often not referable to the syphilitic involvement of the spinal cord The disease is often diagnosed from the investigation which follows the discovery of an Argyll Robertson pupil and absence of knee or ankle tendon reflexes

If the patient seeks medical aid for tabes dorsalis it is often because of disturbances of locomotion or for pain The locomotor disturbances are manifested by disturbances of the sense of position of the parts or losing balance on leaning forward The pain arises from injury by the syphilitic process to the posterior nerve roots and the spinal ganglia which are sensory in function The pain may be mild or severe If mild the pain is described in various ways but generally if severe it is described as sharp boring shooting lightning like lancinating compressing

Muscle tone is increased in disease both of the pyramidal and of the extra pyramidal motor systems but in each case the increase is qualitatively different The spasticity caused by an upper motor lesion is easily to be differentiated from the rigidity of extrapyramidal disease as for instance in Parkinson s disease Both of these types of increased tone are reflex in origin and are abolished if the afferent nerve fiber from the muscle is severed and the reflex arc thus broken Muscle tone is estimated by the manipulation of the limbs to determine the sense of the degree of the active elastic tension in the muscle The knowledge of the normal range of this tension can be gained only through experience

DISEASES MAINLY AFFECTING THE SENSORY NERVOUS SYSTEM

Herpes Zoster (Shingles Zona) Herpes zoster is a specific infectious disease caused by the varicella herpes zoster group of viruses There are a selective inflam mation of the posterior root ganglia and herpetic eruption within the corresponding dermatomes Trauma and diseases and irradiation which involve these ganglia may cause eruptions of the same type and distribution There are therefore a true herpes zoster and a symptomatic type of zoster The vesicles must be regarded as the result of disturbances of function of the ganglia and not as due to the localization of virus in the skin for the disease cannot be transmitted to experimental animals by inoculation with the vesicular fluid It is thought that zoster is related to chicken pox since varicella sometimes develops in persons exposed to zoster and conversely zoster may develop after exposure to varicella

An attack of zoster confers a high grade of immunity However, second attacks do occur

SYMPTOMS This disease occurs sporadically and it is said to occur occasionally in epidemics in summer and autumn It is a weakly contagious disease It is some times possible to trace the infection to a contact

The incubation period is between 1 and 3 weeks The first symptom is pain accompanied with severe cutaneous hyperesthesia The pain corresponds in its dis tribution to one or more dermatomes and may be mild or intense There is often a low fever

The herpes or shingles usually proceed halfway around the body or halfway across the forehead The lesions are characterized by clusters of vesicles on an erythematous base which appear in the region of the skin disturbance of one or more of the posterior spinal nerves The onset of the disease is preceded by or accom panied with neuralgic pain often very severe and tenderness in the part The ery thematous patches more or less oval with the long axis parallel to the underlying nerves occur successively The number of lesions varies from two or three to several dozen Soon the surfaces of the patches are studded with papules which are quickly transformed into vesicles The important feature of the eruption is that it is unilateral Herpes zoster frequently accompanies or is associated with varicella However an attack of varicella does not prevent the patient from having herpes zoster In herpes zoster of the forehead the conjunctiva and the eyeball may be attacked and sometimes the mouth the tongue or the palate may be affected

After 3 or 4 days erythematous patches appear within the area of hyperesthesia Then very quickly small vesicles filled with clear fluid form on these erythematous spots The vesicles dry leaving scars as the result of secondary infection While the disease is acute cervical rigidity and a positive Kernig s sign may be present The spinal fluid often contains in excess of 100 lymphocytes per cubic millimeter

The active phase of the disease lasts only a short time usually a week or less As a rule the pain persists in middle aged and aged patients long after the eruption has healed Hyperesthesia corresponding to the dermatomes involved may persist for years

Shingles rarely appears on the extremities distal to the elbow or the knee The

any time during the course of the disease Ocular palsies resulting in ptosis and strabismus and diplopia are common and are characteristically variable in their course However they may be permanent The other cranial nerves rarely give symptoms Occasionally there is deafness The vagus nerve may be involved resulting in the so-called posticus paralysis a bilateral paralysis of the abductors of the vocal cords

Trophic ulcers perforating ulcers especially of the plantar surface of the toes and balls of the feet are occasionally present These ulcers are very deep and often penetrate to the bones Syphilitic clubfeet as the result of involvement of bones and joints of the feet may be present

Muscular atrophies are rarely encountered Occasionally a foot drop due to atrophy of the peroneal group of muscles and less frequently atrophy of the tongue are present Plantar extension indicates vascular neurosyphilis with involvement of the pyramidal tract or a supervening general paresis otherwise it is not present in tabes dorsalis

In the examination for tabes dorsalis if the disease should be typically manifest the columnae gracilis and cuneatus (Gall and Burdach) are affected These columns lie between the posterior horns of gray matter and the posterior septum They convey the senses of touch position (proprioceptive sense) and vibration However some of the fibers for touch are in the anterior fasciculi If these posterior columns are affected there is a flaccidity of the muscles and the joints and a positive *Romberg sign*

In testing for the Romberg sign request the patient to stand with the feet close together If this is accomplished satisfactorily the examiner stands behind the patient in a position of readiness to catch him if there should be evidence of loss of balance and the patient is asked to close the eyes If there is more than normal swaying when the eyes are closed the sign is positive and there is disease of the posterior columns Aside from the normal uncertainty in standing in labyrinthine and cerebellar disease the Romberg sign is negative in these disorders

Involvement of the sensory nerve roots by the syphilitic process produces objective and subjective sensory disturbances The zones of hyperalgesia and hyperesthesia of radicular distribution corresponding to the level of the nerves affected are frequently situated in the midthoracic and the lower lumbar regions There is absence of nerve trunk tenderness In rare instances the cutaneous distribution of the trigeminus nerve is affected and thus paresthesias are present about the nose and cheeks

The deep sensibility is characteristically affected the change is manifested by loss of sense of position of various members for instance of movements of the toes Early impairment of position and vibratory sensation is always found This may be slight at first or may affect only the toes or vibratory sensation may be lost only over the sacrum In cervical tabes the process begins in the cervical segments so that the disturbance of position sensation and ataxia begin in the upper extremities

The serologic reactions of the blood are positive in from 60 to 75 per cent of those who have tabes The serologic reactions of the spinal fluid generally are positive As there is invariably some degree of meningeal inflammation there usually is an increase of cells in the spinal fluid anywhere from 10 to 100 per cubic millimeter This pleocytosis depends on the extent of the meningeal reaction the acuteness of the process the stage of the disease and the influence of treatment In quiescent forms of the disease there may be no cells in the spinal fluid and serologic reactions may be completely negative The globulin content is usually increased The colloidal gold curve is tabetic

DIAGNOSIS An Argyll Robertson pupil is a sign of parenchymatous neurosyphilis namely tabes and paresis

In paresis an Argyll Robertson pupil is associated with prominent mental symp

or squeezing The attacks of pain are often paroxysmal come on fairly suddenly last a few hours days or weeks, and cease abruptly only to recur again and again Some patients are free from them others are tormented by them throughout the course of the disease The distribution of the pain depends on the situation of the syphilitic process If the lumbosacral roots are affected there are pains in the legs if the lower cervical roots are involved the pain is frequently along the ulnar side of the arm if the lower thoracic roots are affected the pains are in the abdomen The pain may be localized in the thorax or it may affect the trigeminal roots and simulate neuralgia

As the result of syphilitic injury to the sacral paths there are complaints of disturbances in the genital vesical and anal functions Impotence and loss of libido in men and genital anesthesia in women may occur during the course of the disease Paresis of the bladder (cord bladder) manifested by retention or incontinence of urine may be complained of Relaxation of the anal sphincter which may occur early in the disease and anal incompetence with loss of feces are grave symptoms

Affections of the joints of trophic origin (Charcot joints) may occur in tabes These joint disturbances may appear after trauma but because of the absence of pain the patient is unaware of the seriousness of the injury A trophic syphilitic joint never improves the course is progressive Charcot joints as the result of destruction of the vertebrae may permit of serious injury to the spinal cord The symptoms of cord injury are those of a transverse myelitis Tabetic clubfoot may bring the patient to the physician Spontaneous fractures of the bones resulting from the presence of gummas are rare

The impairment of both superficial and deep sensibilities is characteristic of tabes dorsalis Very early in the course of the disease there may be a feeling in the feet as if they were treading on a soft surface such as a carpet or velvet

During the course of tabes dorsalis there are often feelings of *euphoria* The *family may relate incidents and evidences indicating fallacies* of the patient s *judgment*

EXAMINATION Ataxia is often present which depends on impairment or abolition of deep position sensation carried by the posterior columns The ataxia is manifested by a sense of insecurity with uncertainty in walking This uncertainty causes steppage gait in which each foot is raised higher than in normal gait and then is slapped flatly to the ground A tabetic patient would rather ascend a stairway then descend There is great difficulty in walking in the dark There may be bruises of the face or forehead On washing the face if the patient closes the eyes a fall forward and injury by striking the forehead against the wall may occur As the ataxia increases the tabetic patient walks with the feet farther apart in order to broaden the base of support The uncertainty of the gait is further manifested by the close watch the patient keeps on each step and the use of walking canes Muscular hypotonia is present It may be so marked as to give hyperextension at the knees (*genu recurvatum*) and flat feet (*flaccid pes planus*)

Very early in tabes the deep reflexes are diminished and then abolished Loss of knee jerks and ankle jerks is common to all tabetic patients and may even be the first sign and precede the other signs and symptoms by months or years The abdominal reflexes may be hyperactive

The pupillary reflexes are disturbed early The disturbance may originate in the ciliary ganglion or the pathway between it and the primary visual center in the centers themselves in the nuclei of the third nerve or in the sympathetic pathways The pupils may be miotic irregular and unequal They usually do not react to light but do react during accommodation This is the Argyll Robertson pupillary action Once the Argyll Robertson pupil is present it is permanent

The ophthalmologist may be the first to announce the presence of tabes dorsalis The optic nerve is commonly affected Blindness from optic atrophy may result at

Bilateral lesions produce bilateral anesthesia and unilateral lesions cause the Brown Sequard syndrome in which there are analgesia and thermanesthesia on the side opposite to the lesion and loss of proprioceptive sensibility on the side of the lesion If analgesia and thermanesthesia are found over an area corresponding to that of one or more dermatomes with little or no loss of tactile sensibility a lesion in the gray matter of the spinal cord is suspected

Congenital Malformations Involving the Spinal Cord and Meninges ***Diplomyelia*** The term diplomyelia refers to doubling or duplication of the spinal cord The duplication exists in the lower part of the cord and rarely if ever extends above the midthoracic region There is no constant disturbance of function associated with diplomyelia In approximately half of all cases there is spina bifida of the occult variety Other abnormalities are common and usually are grossly manifested

Myelodysplasias *Myelodysplasia with spina bifida* is a defective development of the spinal cord associated with defect in the overlying vertebral laminas

In *complete rachischisis* the neural tube lies open and exposed for a number of segments There is always paralysis

Meningomyelocele is a rounded surface tumor containing the spinal cord which has retained its connection with the surface ectoderm and is attached firmly to the sac wall

Meningocele is a bulging surface tumor covered with normal skin Within the skin is a sac composed of the meninges which may or may not be fused so that the sac has a single or a double wall

In *spina bifida occulta* the laminae of the vertebrae are defective In rare instances the site of the lesion is marked by changes in the overlying skin such as abnormal growth of hair a dimple a deposit of fat or telangiectases In the majority of these persons there are no symptoms In an occasional individual when the posterior surface of the spinal cord is attached to the meninges paralysis is associated In other patients a progressive gliosis may develop and cavities appear in the gray matter so that the symptoms are similar to those of syringomyelia

There is a malformation of the cervical spine in which the vertebrae are reduced in number and fused into one mass of bone Spina bifida is usually present This condition is termed *myelodysplasia with Klippel Feil syndrome*

Myelodysplasia may be associated with *absence of the sacrum and coccyx* The normal prominences of the buttocks are lost and the intergluteal fold is also absent Palpation reveals soft tissue where the sacrum should be Roentgenologic examination reveals the defect (Arey)

Myelitis and Abscess of the Spinal Cord The staphylococci and streptococci commonly but occasionally other pyogenic organisms may reach the spinal cord by way of the blood stream by penetrating wounds or by direct extension from adjacent foci of infection and thus originate a myelitis or an abscess

The onset of the myelitis is often obscured by the presence of symptoms due to the primary infection It usually requires a severe pain in the back and the rapid development of a flaccid paralysis of the legs in order that the symptoms be noticeable or attributable to disease of the spinal cord In some patients the retention of urine is first observed On examination the tendon and the plantar responses are abolished Sensibility is lost below the level of the lesion In some instances there has been no serious illness The history reveals that there was a skin infection or trauma with infection which seemingly healed After this the symptoms of a spinal cord tumor developed

The spinal fluid contains a variable number of leukocytes and lymphocytes depending on the degree of meningeal involvement Protein is almost always increased in the spinal fluid

Once the symptoms of myelitis and abscess of the cord are established bed sores develop despite the greatest care and death usually ensues from paralysis of respiration meningitis or sepsis if antibiotic therapy is ineffective

The diagnosis is made by recognizing a transverse myelitis of rapid onset developing in association with septicemia or with a suppurative process

toms Other forms of neurosyphilis usually show a fixed pupil and are characterized by signs of disseminated lesions which come on much earlier in the course of syphilis are much more acute in their onset and also respond better to treatment The Argyll Robertson pupil in a patient who has a history of primary or secondary syphilis even in the presence of no other manifest symptoms is almost diagnostic of tabes The Argyll Robertson pupil in association with a history of pains in various radicular zones or a diminished or absent knee jerk or ankle jerk or both or a history of girdle sensation and vomiting or the presence of ataxia or a history of vesical disturbance with impotence or disturbed position and vibratory sensation in the toes and hyperesthesia over the nipples buttocks and ankles or loss of nerve tenderness or early optic atrophy—any combination of these—is sufficient to establish a diagnosis of tabes dorsalis Tabes dorsalis may be diagnosed in the absence of pupillary signs provided there are signs and symptoms indicative of involvement of posterior roots and posterior columns accompanied with a positive serologic reaction for syphilis These symptoms and signs are pains crises loss of deep reflexes and ataxia The presence of positive or of negative serologic reactions of the blood may influence an error in diagnosis A positive serologic reaction of the blood alone is not diagnostic A positive serologic reaction for syphilis in the spinal fluid is diagnostic if accompanied by appropriate signs and symptoms

Differentiation of tabes dorsalis may be extensive because of the many possible manifestations of the disease Some diseases to be differentiated are multiple neuritis (polyneuritis) combined sclerosis of the cord and Adie s syndrome

The etiology and pathogenesis of Adie s syndrome are unknown The association of this syndrome with other disorders such as influenza polyneuritis encephalitis multiple sclerosis orthostatic hypotension vitamin deficiencies and endocrine disorders particularly hypothyroidism has been suggested However the syndrome seems to be more frequently associated with nervous and emotional disorders The syndrome is characterized by tonic pupils and sometimes absent tendon reflexes

A tonic pupil is one having a delayed or slow response to accommodation and convergence The pupil constricts more than the normal pupil under the same stimulus It responds slowly to bright light and dilates in the same manner after the stimulus has been withdrawn The tonic pupil is usually unilateral although it may be bilateral The abnormal pupil is usually larger than the normal pupil The tonic pupil reacts normally to both atropine and physostigmine The Argyll Robertson pupils on the other hand are usually miotic bilateral and fixed to light stimulation but react promptly to convergence

The tendon reflexes most frequently absent in Adie s syndrome are the Achilles and patellar reflexes although all tendon reflexes may be absent In contrast to tabes dorsalis the patient with Adie s syndrome has no loss or disturbance of vibratory or position sense and none of the superficial sensory disturbances lightning pains or gastric crises

There are variations in the findings in Adie s syndrome Some patients have tonic pupils alone and others have absence of reflexes alone In the intermediate positions are those having atypical pupils with absence of one or more tendon reflexes

DISEASES OF THE SPINAL CORD

In association with the brain and its meninges the spinal cord and its meninges share in a number of infections due to bacteria viruses and higher plant and higher animal parasites Here will be considered some of the diseases which often profoundly affect locomotion namely some of the more chronic infections acute injuries and metabolic vascular unknown and tumorous diseases of the cord and its covering

Loss of sensibility on one or both sides of the body up to a level corresponding to the lower border of a dermatome indicates that a lesion is in the spinal cord

and impotence are frequent and persistent There may be ataxia or chronic progressive spasticity

There may be present pupillary signs most often fixed both to light and on accommodation Ocular palsies are common If the process be limited to the region about the central canal there may be a dissociation of pain and temperature sense as in syringomyelia

DIAGNOSIS A history of syphilis positive serologic reactions in the presence of signs of meningeal reactions and cord changes an increased number of cells in the spinal fluid increased globulin and a positive colloidal gold curve are diagnostic of meningomyelitis The serologic reaction of the spinal fluid may be negative

The outlook in syphilitic meningomyelitis depends on the duration of the process on the extent of involvement and on the treatment The disease is not cured but symptomatically the patient may improve under proper treatment

Traumatic Lesions of the Spinal Cord As the result of trauma the spinal cord may be injured without fracture or dislocation of the spinal column The history of an injury to the vertebrae signifies the nature of the injury

The roentgenologic examination will reveal fractured or dislocated vertebrae if they are present In rare instances however no lesions of bone may be demonstrable roentgenologically The anatomic character and severity of the lesion in the spinal cord frequently cannot be determined at once If the paralysis is not complete at first the injury is relatively mild but if there is complete loss of conduction in the cord it is impossible to say whether the cord is actually transected or whether there is a transient loss of function without complete anatomic dissolution

A sudden total transection of the spinal cord causes immediately total flaccid paralysis of the limbs sensory loss loss of tendon jerks retention of urine and sometimes transient absence of all plantar response There is spinal shock If the patient does not die from infected bedsores or cystitis spinal shock passes off in a few weeks The knee and ankle jerks may return but the plantar response remains extensor Stroking of the sole may produce involuntary passage of urine

When the lesion is incomplete the reflexes return after a relatively short time and the muscle tone returns chiefly in the extensor If signs of compression of the spinal cord develop weeks or months after an injury of the back the following possibilities should be considered delayed traumatic spondylopathy associated with deformity of the spinal column due to collapse of a vertebra osteomyelitis tuberculous spondylitis and above all the extrusion of an intervertebral disk

Death in the first few hours may occur from paralysis of respiration from shock or from associated injuries After the first few days the danger is chiefly from infected decubitus ulcers infections of the urinary tract and pneumonia Lesions high enough to weaken the muscles of respiration cause a high mortality rate

The prognosis as regards recovery depends on the severity of the injury If the lesion is only partial there is always much functional improvement

Kuhn and Macht have observed the involuntary activity patterns of skeletal muscles innervated below the level of severance in 27 instances of complete section of the spinal cord Some patients survived as long as 42 months The progression of reflex activity following spinal cord transection in this series of patients was spinal shock minimal reflex activity flexor spasms alternating flexor and extensor spasms and predominant extensor spasms The length of time that different subjects remain in a given stage varies considerably In general stage 1 lasts for 1 to 6 weeks stages 2 and 3 6 weeks to 1 year stage 4 rarely begins prior to 4 months after injury and usually is terminated within 1 year and stage 5 may appear as early as 6 months after the injury and may continue indefinitely Extensor thrust is not common but can be elicited occasionally in spinal man Spinal standing similar to that seen in animals with transected cords can occur in spinal man Sustained patellar and ankle clonus frequently occurs in chronic spinal man Primary extensor

Pyogenic myelitis is almost invariably fatal However drainage of such an abscess by Woltman and Adson led to recovery

Pyogenic Infections of the Spinal Epidural Space Furunculosis caused by staphylococci is the commonest cause of infection of the epidural space There is fever as well as severe pain in the back which extends along the course of the spinal nerve roots Flaccid paraplegia develops in about one week There are rigidity of the spinal column and local tenderness The spinal fluid contains an excess of cells Pus may be obtained from the epidural space when lumbar puncture is attempted

The diagnosis of extradural abscess depends on the rapid development of para plegia which usually ascends on the rigidity of the spinal column on the signs of an acute septic infection and most important of all on the discovery of pus in the epidural space

The prognosis in cases of extradural abscess is very poor Antibiotic therapy is used

Hemorrhagic Myelitis Hemorrhagic myelitis is a secondary process due to various bacterial viral and other poisons Any acute infectious disease may cause a hemorrhagic myelitis for instance scarlet fever diphtheria rickettsial disease ma laria pneumonia and septicemia to mention only a few

The symptoms are those of transverse myelitis disseminated myelitis or ascend ing myelitis with flaccid paralysis The tendon reflexes are lost the plantar response is extensor and there is retention of urine The paralysis of some of the muscles is permanent The disturbance of sensibility if present disappears within a short time

The mortality rate is high and averages between 30 and 50 per cent Death generally occurs between the fifteenth and eighteenth days and is associated with coma and bulbar palsy bronchopneumonia and infection of the urinary tract Some of those who have the disease recover while others sustain a lasting disability

Syphilitic Meningomyelitis *Neurosyphilis produces syndromes which refer* to the spinal cord the brain or the meninges In each of these syndromes there are symptoms which indicate an obvious fact that the entire nervous system is affected in neurosyphilis

The changes caused by syphilis may be largely limited to the spinal cord alone or to the spinal meninges generally it involves both The infection very often begins in the meninges There are inflammation and fibrous thickening of the meninges and adhesions between the dura and piarachnoid which may contain granulations or gum matous tumors There are endarteritic changes strangulated thrombosed vessels and hemorrhage with secondary spinal softening and degeneration The inflammatory process may extend to the roots of the spinal nerves causing atrophy of the nerve fibers When the syphilitic process affects mainly the spinal cord there are softening and atrophy In the cord the process may be selective and remain largely restricted to the periphery limited to certain columns and tracts or it is partial or complete at only one level of the cord In other instances of cord involvement the process is diffuse and disseminated

Symptoms The meningovascular variety of neurosyphilis commences within the first 5 or 6 years of the infection There may be paraplegia spinal hemiplegia quadriplegia or a Brown Sequard syndrome The paralysis is generally spastic but may be flaccid The syndrome may simulate that of subacute poliomyelitis with segmental atrophies and no sensory disturbances

There are often pains in the root zones These pains are present in the neck back or loins The pains are often girdle like and follow definite paths along the arms or legs and whether mild or severe they are worse at night than in the daytime Spasms and atrophies of the muscles are present when the motor roots are affected Meningomyelitis occasionally is manifested by pains ataxia and vesical disturbances and severe headaches are common

Examination There is a loss of tendon reflexes Patchy sensory disturbances a loss or diminution of sensation below the level of the lesion in the paraplegias spinal hemiplegias, and quadriplegia are common Vesical and rectal disturbances

the preservation of the axis cylinder accounts for the possibility of remissions It is only when both the myelin sheath and the axis cylinder are destroyed that the signs and symptoms become permanent

SYMPTOMS Multiple sclerosis may be manifested by disturbed functions in one or many different parts of the nervous system For instance neurologists distinguish a *sacral* form of the disease characterized by sphincteric disturbances of the anus and bladder The *cervical* form of the disease is manifested by acute unilateral ataxia astereognosis slight spasticity and sensory changes In the *bulbar* form the process is characterized by disturbance in phonation articulation and deglutition If there are disturbances of function of the facial and trigeminal nerves the disease is designated as *pontine* multiple sclerosis Asynergy characterizes a *cerebellar* form of multiple sclerosis *Cortical* forms of the disease are rare but when present the manifestations are acute hemiplegia and ophthalmoplegia There are however a number of signs and symptoms which occur with sufficient frequency and consistency to make multiple sclerosis a unified clinical disease

A sufficient number of patients who have multiple sclerosis have scanning speech tremor and nystagmus so that these constitute a triad of symptoms considered to be diagnostic of the disease However these are but a few of the symptoms usually present in the disease

The first symptoms such as a fleeting ocular palsy manifested by temporary strabismus or a slight visual disturbance or a mild leakage of urine or a transient weakness of a limb often signify the onset of multiple sclerosis months or years before the patient is aware that a serious disease is developing

A disturbance of locomotion manifested by difficulty in walking often is the first definite symptom The gait becomes spastic or ataxic and there is increasing hypertonicity of the lower limbs Often the patient stumbles or falls in walking The greatest in-co ordination is often in the upper extremities Acute ataxia of an arm alone may occur

In rare instances the onset is acute and is manifested by apoplexy The syndrome is ushered in by hemiplegia at times accompanied by unconsciousness and rarely even by a convulsion In rare instances too the illness is that of a transverse myelitis in these however the acute symptoms usually recede

A slowing halting scanning or explosive speech is a common symptom of multiple sclerosis The words are broken up into syllables and scanned a form of articulation which is in reality a dysarthria Scanning is extremely characteristic

There may be difficult micturition or slight transitory incontinence Vesical incontinence occurs when there is severe involvement of the sacral segment of the spinal cord Anal incontinence is rarely present

Emotional disturbances such as an impulsive laughter more rarely crying often occur early

EXAMINATION In the early stages of the disease in the interim between attacks results of examination may be negative

Coincident with or preceding the development of spasticity the knee jerks and ankle jerks become hyperactive Plantar extension and clonus are present (Babinski sign) There are early impairment and loss of the abdominal reflexes Loss of abdominal reflexes accompanied with an ataxic tremor with an intention element are pathognomonic signs of the disease The tremor comes on gradually usually in the upper extremities and frequently becomes extreme It may become so disturbing as to interfere with dressing and eating The tremor usually disappears at rest and is absent during sleep

Lateral nystagmus is an early and common sign of multiple sclerosis It indicates a focus in the medulla or pons

There may be areas of hypalgesia and hypesthesia or even complete loss of

manifestations in chronic spinal man probably represent the reappearance of primitive and poorly integrated postural activity mediated by the isolated cord the functional capacity of which seems greater than has been assumed Although spinal man passes through a period in which flexion reflexes alone are active he may frequently progress to a stage of activity characterized by predominantly extensor reflexes amounting in many cases to extensor spasm It is probable that many of the discrepancies between these observations and those of earlier investigators can be explained by the better physical conditions of present day spinal men

Circulatory Diseases of the Spinal Cord The intraspinal blood supply is derived from the central or anterior sulcal branch of the anterior arterial trunk and the peripheral arteries arising from the pial plexuses These two systems although possessing anastomosis of capillaries function as end arteries *The arterial distribution is not according* to anatomic or physiologic relations The larger vessels pass into the cord and supply the ganglion cells

The veins emerging from the interior of the spinal cord are united into extensive venous plexuses on the surface upon which are situated the main longitudinal channels

Occlusion of the Anterior Spinal Artery When the anterior spinal artery is occluded, there are sudden paraplegia absence of defense reactions dissociated pain and temperature sensory disturbances accompanying disturbances of vesical and intestinal functions These symptoms vary according to the level involved Flaccid muscle paralysis and segmental atrophy ensue

Atherosclerosis Atherosclerotic changes in the blood vessels may be present in the spinal cord and are most intense in the thoracic part of the cord Likewise in syphilis the upper thoracic part of the cord will be most affected by syphilitic arteritis

Mild but distinct evidences of degeneration of the posterior column and horn appear in vascular disorders of old age

In generalized atherosclerosis signs of defective nutrition of the cord appear under stress or exertion similar to what occurs in the brain Like that of the brain the anemia of the cord is *intermittent in character*

The most frequent symptom is numbness and a feeling of coldness in the lower extremities There is a diminution or loss of vibratory and joint sensibility and sometimes the sense of touch is impaired There may be local muscular atrophies and spasticity

The effect of atherosclerotic changes in the vessels supplying the peripheral nerves is to decrease the blood supply to the peripheral nerves as well as to the other tissues There results the so called arteriosclerotic neuritis The symptoms consist of marked pain hyperesthesia of the skin atrophy and glossiness of the skin and occasionally some objective sensory changes and muscular atrophy

Disseminated (Multiple) Sclerosis Multiple sclerosis is a *chronic* intermittent disease of the central nervous system Remission of a part or all of the symptoms may be short or may last over a period of years Usually however there are repeated relapses with steady aggravation and progression of all signs and symptoms until invalidism and death result

Multiple sclerosis is essentially a disease of early adult life in most cases occurring between the ages of 20 and 40 years The disease is commoner in men *than in women*

The constant pathologic features of multiple sclerosis consist mainly of numerous and widespread foci of sclerosis of varying ages of duration throughout the central nervous system degeneration of the myelin sheath and preservation of the axis cylinder *in the midst* of the foci of sclerosis (periaxial neuritis)

The sclerotic process is situated more in the white matter of the spinal cord especially the pyramidal tracts and in the structures of the medulla pons midbrain and the optic chiasm and tract Owing to the varying ages of the *patches of the* sclerosis the patient may be observed to have some *signs and symptoms which* remain permanent and others which are capable of remission

Degeneration of the axis cylinder proceeds until it is finally destroyed The destruction *of the myelin* sheath alone permits of only a partial impairment of function while

the preservation of the axis cylinder accounts for the possibility of remissions It is only when both the myelin sheath and the axis cylinder are destroyed that the signs and symptoms become permanent

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There may be areas of hypalgesia and hypesthesia or even complete loss of

manifestations in chronic spinal man probably represent the reappearance of primitive and poorly integrated postural activity mediated by the isolated cord the functional capacity of which seems greater than has been assumed Although spinal man passes through a period in which flexion reflexes alone are active he may frequently progress to a stage of activity characterized by predominantly extensor reflexes amounting in many cases to extensor spasm It is probable that many of the discrepancies between these observations and those of earlier investigators can be explained by the better physical conditions of present day spinal men

Circulatory Diseases of the Spinal Cord The intraspinal blood supply is derived from the central or anterior sulcal branch of the anterior arterial trunk and the peripheral arteries arising from the pial plexuses These two systems although possessing anastomosis of capillaries function as end arteries The arterial distribution is not according to anatomic or physiologic relations The larger vessels pass into the cord and supply the ganglion cells

The veins emerging from the interior of the spinal cord are united into extensive venous plexuses on the surface upon which are situated the main longitudinal channels

Occlusion of the Anterior Spinal Artery When the anterior spinal artery is occluded there are sudden paraplegia absence of defense reactions dissociated pain and temperature sensory disturbances accompanying disturbances of vesical and intestinal functions These symptoms vary according to the level involved Flaccid muscle paralysis and segmental atrophy ensue

Atherosclerosis Atherosclerotic changes in the blood vessels may be present in the spinal cord and are most intense in the thoracic part of the cord Likewise in syphilis the upper thoracic part of the cord will be most affected by syphilitic arteritis

Mild but distinct evidences of degeneration of the posterior column and horn appear in vascular disorders of old age

In generalized atherosclerosis signs of defective nutrition of the cord appear under stress or exertion similar to what occurs in the brain Like that of the brain the anemia of the cord is intermittent in character

The most frequent symptom is numbness and a feeling of coldness in the lower extremities There is a diminution or loss of vibratory and joint sensibility and sometimes the sense of touch is impaired There may be local muscular atrophies and spasticity

The effect of atherosclerotic changes in the vessels supplying the peripheral nerves is to decrease the blood supply to the peripheral nerves as well as to the other tissues There results the so called arteriosclerotic neuritis The symptoms consist of marked pain hyperesthesia of the skin atrophy and glossiness of the skin and occasionally some objective sensory changes and muscular atrophy

Disseminated (Multiple) Sclerosis Multiple sclerosis is a chronic intermittent disease of the central nervous system Remission of a part or all of the symptoms may be short or may last over a period of years Usually however there are repeated relapses with steady aggravation and progression of all signs and symptoms until invalidism and death result

Multiple sclerosis is essentially a disease of early adult life in most cases occurring between the ages of 20 and 40 years The disease is commoner in men than in women

The constant pathologic features of multiple sclerosis consist mainly of numerous and widespread foci of sclerosis of varying ages of duration throughout the central nervous system degeneration of the myelin sheath and preservation of the axis cylinder in the midst of the foci of sclerosis (periaxial neuritis)

The sclerotic process is situated more in the white matter of the spinal cord especially the pyramidal tracts and in the structures of the medulla pons midbrain and the optic chiasm and tract Owing to the varying ages of the patches of the sclerosis the patient may be observed to have some signs and symptoms which remain permanent and others which are capable of remission

Degeneration of the axis cylinder proceeds until it is finally destroyed The destruction of the myelin sheath alone permits of only a partial impairment of function while

the preservation of the axis cylinder accounts for the possibility of remissions It is only when both the myelin sheath and the axis cylinder are destroyed that the signs and symptoms become permanent

SYMPTOMS Multiple sclerosis may be manifested by disturbed functions in one or many different parts of the nervous system For instance neurologists distinguish a *sacral* form of the disease characterized by sphincteric disturbances of the anus and bladder The *cervical* form of the disease is manifested by acute unilateral ataxia astereognosis slight spasticity and sensory changes In the *bulbar* form the process is characterized by disturbance in phonation articulation and deglutition If there are disturbances of function of the facial and trigeminal nerves the disease is designated as *pontine multiple sclerosis* *Asynergy characterizes a cerebellar form* of multiple sclerosis *Cortical* forms of the disease are rare but when present the manifestations are acute hemiplegia and ophthalmoplegia There are however a number of signs and symptoms which occur with sufficient frequency and consistency to make multiple sclerosis a unified clinical disease

A sufficient number of patients who have multiple sclerosis have scanning speech tremor and nystagmus so that these constitute a triad of symptoms considered to be diagnostic of the disease However these are but a few of the symptoms usually present in the disease

The first symptoms such as a fleeting ocular palsy manifested by temporary strabismus or a slight visual disturbance or a mild leakage of urine or a transient weakness of a limb often signify the onset of multiple sclerosis months or years before the patient is aware that a serious disease is developing

A disturbance of locomotion manifested by difficulty in walking often is the first definite symptom The gait becomes spastic or ataxic and there is increasing hypertonicity of the lower limbs Often the patient stumbles or falls in walking The greatest in-co ordination is often in the upper extremities Acute ataxia of an arm alone may occur

In rare instances the onset is acute and is manifested by apoplexy The syndrome is ushered in by hemiplegia at times accompanied by unconsciousness and rarely even by a convulsion In rare instances too the illness is that of a transverse myelitis in these however the acute symptoms usually recede

A slowing halting scanning or explosive speech is a common symptom of multiple sclerosis The words are broken up into syllables and scanned a form of articulation which is in reality a dysarthria Scanning is extremely characteristic

There may be difficult micturition or slight transitory incontinence Vesical incontinence occurs when there is severe involvement of the sacral segment of the spinal cord Anal incontinence is rarely present

Emotional disturbances such as an impulsive laughter more rarely crying often occur early

EXAMINATION In the early stages of the disease in the interim between attacks results of examination may be negative

Coincident with or preceding the development of spasticity the knee jerks and ankle jerks become hyperactive Plantar extension and clonus are present (Babinski sign) There are early impairment and loss of the abdominal reflexes Loss of abdominal reflexes accompanied with an ataxic tremor with an intention element are pathognomonic signs of the disease The tremor comes on gradually usually in the upper extremities and frequently becomes extreme It may become so disturbing as to interfere with dressing and eating The tremor usually disappears at rest and is absent during sleep

Lateral nystagmus is an early and common sign of multiple sclerosis It indicates a focus in the medulla or pons

There may be areas of hypalgesia and hypesthesia or even complete loss of

sensation Disturbance of deep sensation such as position and vibration are found if the disease affects the posterior columns Astereognosis may be profound

Ophthalmologic examination often reveals a bitemporal pallor of the optic disks and central scotomas Optic atrophy may produce blindness *However all of these* visual disturbances of the eyes including total blindness may show remissions

Ocular palsies with strabismus and double vision occur early or at any time during the course of the disease Any disturbance of pupillary reactions however *is not due to multiple sclerosis* Internal ophthalmoplegias are rare in multiple sclerosis

Bulbar and cranial nerve manifestations are rarely present Occasionally paralysis of the vocal cords disturbances of phonation chewing and swallowing and facial paralysis and deafness may occur

The spinal fluid in multiple sclerosis may show a slight lymphocytosis and a paretic gold curve The spinal fluid pressure and globulin concentration are within normal limits The serologic reactions *are negative*

DIAGNOSIS Increased tendon reflexes spasticity of the lower extremities plantar extension (Babinski sign) tremor nystagmus speech disturbance ataxia impulsive laughter central scotomas bitemporal pallor and negative serologic reactions in a patient who has had these symptoms remittently for a long time are diagnostic of multiple sclerosis

Combined Sclerosis (Ataxic Paraplegia) Combined sclerosis is a progressive disease of the spinal cord which most frequently results from pernicious anemia The disease may appear in the course of pernicious anemia in the absence of severe anemia It may advance to a paraplegic state

There is degeneration of the posterior and lateral columns of the spinal cord *The disease often commences with paresthesias which are prominent through*out its course Involvement of the posterior columns gives varying degrees of ataxia of the lower extremities and occasionally of the arms The legs are almost always involved first There is frequently pseudoathetosis of the fingers

The tendon reflexes are often absent and vibration and position sensation are impaired or lost In instances of degeneration of the pyramidal tract there may be hypertonia with increased reflexes and finally a spastic ataxic paraplegia The sensory disturbances may affect all forms of sensation Incontinence of urine and bedsores attest the severity of the cord involvement late in the course of the disease

The onset of paresthesias ataxia disturbed vibratory and position sensation diminished deep reflexes and the presence of plantar extension and paraplegia *accompanied with pernicious anemia are diagnostic* Pellagra may give an ataxic paraplegia but this is a true myelitis

Syringomyelia (Spinal Gliosis) Syringomyelia is thought to be the result of an embryonic defect of development Under normal conditions the primitive glial cells *migrate peripherally from the vicinity of the central* canal of the spinal cord and differentiate into adult glial cells Probably in syringomyelia spongioblasts fail to migrate and eventually proliferate and thus form dense regions of gliosis in the central portions of the cord which later turn into cavities One or more cavities are present *in the region of the cervical* enlargement but may be present or extend to any segment of the spinal cord Occasionally the process begins in the lumbosacral enlargement rather than in the cervical region The cavities from their points of origin in regions of gliosis in the gray matter near the central canal invade the anterior gray matter and the lateral columns or compress these structures

When a syrinx invades the medulla oblongata (syringobulbia) there is a narrow slit like cavity extending laterally and anteriorly from the fourth ventricle and involving the nucleus ambiguus the hypoglossal nucleus the descending root of the fifth nerve and perhaps the tractus solitarius Median slits are rare

Men and women past the age of 20 years are affected with equal frequency *It is very* rare for more than one instance of the disease to appear in a family

SYMPTOMS The patient may complain of having burnt a foot or a hand in water too hot for bathing without knowing it until after the burn was sustained Some muscles or groups of muscles may twitch and may have become weak shrunken or perhaps paralyzed The fascicular twitchings of the muscles progress up the arm as the muscles become weak and shrink If the symptoms of weakness and shrinkage of the arms have been present for some time paraplegia may have occurred and the patient is confined to the bed and to the wheel chair

Pain is not a common symptom in syringomyelia Occasionally however there may be severe localized pains which involve one or both lower extremities In some the pain is due to the scoliosis and is relieved by supporting the spinal column

EXAMINATION On examination many and diverse findings are likely to be present which are widely scattered and which may be difficult to classify The immediately noticeable abnormalities are atrophy and fibrillary twitchings of the affected muscles deformities of the fingers and hands and perhaps scoliosis in the thoracic region of the spinal column The gait is unsteady and station is uncertain There is paralysis in association with atrophy of the muscles with loss of reflexes and flaccidity which usually occur first in the hands or arms Involvement of the intrinsic muscles of the hands causes various deformities of the fingers After the fingers are involved atrophy affects successively the muscles of the forearm arm and the shoulder girdle The muscles of the neck spinal column and thorax become atrophic toward the end of life Fascicular twitchings in the affected muscles may be present The muscular atrophy is not symmetric In advanced instances of the disease there are paraplegia and spasticity of the legs There is a loss of sphincteral control

In some instances changes will be present in the joints of the upper extremities which are identical with the Charcot joints of tabes dorsalis There are destruction of the cartilage and articular surfaces and relaxation of the capsule and ligaments so that an abnormal range of movement is possible The bones may also become decalcified causing spontaneous fractures Local overgrowth of bones may occur and the hand may resemble that of acromegaly

When the lumbosacral part of the spinal cord is the situation of the syrinx the atrophy and weakness begin in the feet calves thighs or hips Irregularities of sphincteral control in these patients are an early symptom

There are changes in the skin and soft tissues The skin may be dry and thick or thin and glossy The thick nails may become opaque and shed Cyanosis edema and ulcerations may occur on the fingers causing extensive loss of tissue

There is a loss of pain sense tactile sense is not seriously disturbed In some instances all modalities of cutaneous sensibility may be lost in certain regions The distribution of the anesthesia is variable It may be bilateral as the result of interruption of the dorsal commissure It may be unilateral as the result of interruption of the posterior horn of gray matter Sometimes the anesthesia involves the hand or the hand and forearm presenting the so-called glove or sleeve anesthesia In most instances the anesthesia is confined to the arms shoulders neck and thorax As the process in the cord advances and the posterior columns of the cord are affected ataxia loss of proprioceptive sensibility unsteadiness of station and gait and finally complete anesthesia may develop over the lower half of the body

When the disease involves the cervical enlargements from extension from either side there are often signs of involvement of the cervical sympathetic nerve such as Horner s syndrome or unilateral sweating of the face The descending root of the fifth nerve may be affected causing analgesia and thermanesthesia of the face

Nystagmus is an indication of invasion of the medulla The tongue becomes atrophied and paralyzed on one or both sides The soft palate and pharynx and the vocal cords are often paralyzed The sixth nerve may be involved When a syrinx from the medulla reaches the cervical segments there is paralysis of the sterno

cleidomastoid and trapezii muscles, owing to invasion of the spinal nucleus of the eleventh nerve

DIAGNOSIS The diagnosis depends on the presence of a thermo analgesia a slowly progressive atrophy of the muscles supplied by the cervical enlargement accompanied with dissociated anesthesia in the cervical dermatomes the scoliosis and the trophic changes The presence of a syrinx in the lumbosacral enlargement causes identical signs in the corresponding segments

The differential diagnosis consists of the exclusion of progressive spinal muscular atrophy leprosy Raynaud s disease cervical ribs with compression of the cervical nerve roots and neurosyphilis

Syringomyelia is sometimes accompanied by cervical ribs and if the symptoms happen to be confined to the hand the diagnosis may be in doubt until the extension of the symptoms is sufficient to make the nature of the process obvious

The disease may progress so slowly that the patient may be active for many years As a rule however syringobulbia pursues a relatively short course In all instances the prognosis is ultimately grave

Tumors of the Spinal Cord Tumors of the spinal cord cause locomotor difficulties by compression of the cord Compression of the cord aside from that due to trauma may arise from a decrease in the size of the bony conduit through which the cord passes for instance (1) from lesions of the vertebrae (2) from space occupying lesions of the meninges or nerve roots and (3) from tumors arising within the substance of the cord itself

In lesions of the spinal cord the attention is directed to the symptoms and the findings which help to determine the segmental level of the lesion and the structures involved in the transverse direction at the level affected When a disease is due to involvement of a neuron system it is designated as a simple systemic disease When a disease affects more than one neuron system as in amyotrophic lateral sclerosis it is designated a combined neuron disease

In degeneration of the intramedullary continuation of the posterior roots of the spinal nerves as occurs in tabes dorsalis the cutaneous sensibility and the sensibility of the bones joints fascia and muscles are disturbed Anesthesia for all qualities of sensation or elective losses of one or more sensations may be present The reflexes are lost There is in co ordination of movement of the muscles but muscle power is not diminished The function of the bladder may be impaired or lost

Diseases of the sensory neurons of the cord are distinguished from lesions arising in the peripheral nerves by topography of the sensory disturbance in the peripheral nerve lesions which corresponds to the domain supplied by the peripheral nerve rather than to a distribution corresponding to the domain supplied by a posterior nerve root in a cord lesion

It must be recognized that knowledge alone of the motor sensory reflex and visceral representation in the segments of the cord does not permit diagnosis of focal localization or situation of a gross level lesion in the spinal cord Experience is required in order to evaluate and correlate findings with such facts as for example that a peripheral motor nerve innervating a given muscle rarely if ever arises from a single segment but through the mediation of plexuses has its origin in a combination of fibers from the anterior roots of spinal nerves That is each muscle has a plurisegmental innervation and the innervation of muscles of the extremities is by nerve segments at a higher level

Generally extradural sarcomas are most frequent in childhood Intradural meningiomas are commonest among adults Gliomas of the cord are relatively common Kernohan considers that gliomas of the spinal cord are the same as the cerebral gliomas He describes ependymomas spongioblastomas astroblastomas medulloblastomas and ganglioneuromas They usually arise in the central gray matter and extend over a number of segments of the cord

SYMPTOMS Tumor of the spinal cord commences with *pain localized and persistent* associated with cutaneous hyperesthesia at the level of the tumor The pain is unilateral at first but may become bilateral or girdle in distribution It is precipitated by coughing sneezing and lifting If the cauda equina is involved pain and muscle spasm on flexing the back are severe Often the pain becomes much worse shortly after the patient falls asleep in the reclining position In children night crying after going to sleep is frequent In adults pain disturbs or prevents sleep

EXAMINATION *The findings on physical examination depend on the stage of* development and the situation of the tumor There may be either spasticity or flaccidity Flaccid paralysis develops when the anterior surface of the cord is compressed In these lesions an ataxic gait and a positive Romberg sign soon follow Cutaneous anesthesia below the level of the lesion is not long delayed after loss of proprioceptive sensibility The anesthesia may begin in the dermatomes just below the level of the lesion and spread down but more commonly it begins in the lower part of the body and extends slowly upward until it reaches the level of the pain All modalities of sensibility are affected but unequally on the two sides of the body A more severe anesthesia is likely to be on the side opposite the weaker leg

A lesion situated cephalad to the cervical enlargement of the spinal cord is manifested by muscular hypertonus including the diaphragm and the intercostal muscles Paralysis of respiration may occur Lesions situated in the cervical enlargement cause paralysis of the arms thoracic muscles and legs but not the diaphragm Lesions situated cephalad to the first thoracic segment may cause Horner s syndrome The arms and the legs are paralyzed Tumors of the thoracic segments of the cord cause spasticity and paralysis of the legs Tumors between the thoracic segments of the cord and the lumbosacral enlargement cause spasticity and weakness of the legs One leg is affected more than the other Only the legs and the sphincters are affected when the lesion is situated in the lumbosacral enlargement

A tumor of the cauda equina in some patients causes severe pain which extends along the course of the sciatic nerves The pain is increased by flexion or extension of the spinal column coughing sneezing straining and by certain postures The patient may be comfortable when standing but may have intense pain when lying or sitting Loss of control of the bladder develops early The paralysis is flaccid

There are other patients who have a tumor of the cauda who only have pain and spasm of the flexor muscles of the knee hip and lumbar portion of the spinal column The lumbar lordosis is reduced and the patient walks with the knees semi flexed Anesthesia is present over the buttocks and the posterior surface of the thighs and legs

In all spinal cord tumors pains in the body below the level of the lesion are usually absent Burning tingling or hot and cold sensations are common By the time definite anesthesia is present there is likely to be difficulty in passing urine A loss of sphincteral control is a late symptom of compression of the spinal cord *Loss of control of the bladder occurs before rectal incontinence* Toward the end the legs become spastic and are strongly extended a state which terminates in paraplegia in extension followed by paraplegia in flexion Trophic changes in the skin and decubitus ulcerations may be present

Before any fluid is removed the *intraspinal pressure* is estimated it normally registers between 12 and 15 cm of water As soon as the pressure has been estimated jugular pressure is applied Sudden rise and rapid fall of the fluid on compression of both internal jugular veins indicate free flow of cerebrospinal fluid within the subarachnoid space Slow rise and fall of fluid or its failure to rise on compression of the jugular veins suggests partial or complete intraspinal block

Inability to obtain fluid at the fourth lumbar interspace usually signifies that the tip of the needle has failed to enter the subarachnoid space that fluid is absent or that there is a tumor at this level

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In consideration of the symptoms of mononeuritis it is realized that most peripheral nerves are mixed nerves that is they contain both sensory and motor fibers The interruption by disease or section by trauma of a peripheral nerve causes paralysis of the muscles innervated and also sensory loss or changes in the skin innervated by the same nerve In disease rarely is all of the nerve destroyed when some parts of the nerve are still intact there are manifestations of irritative phenomena in addition to aberrations of sensory function Motor irritative phenomena are spasms of the muscle first small areas of the muscle which are termed fascicular or fibrillar twitchings and later contractures of the paralyzed muscle In about three weeks atrophy of the flaccidly paralyzed muscle ensues

The sensory irritative phenomena are mainly pain limited to the distribution of the affected nerve and described as hot burning scalding throbbing or shooting in character Paresthesia dysesthesia causalgia and tenderness along the nerve are common

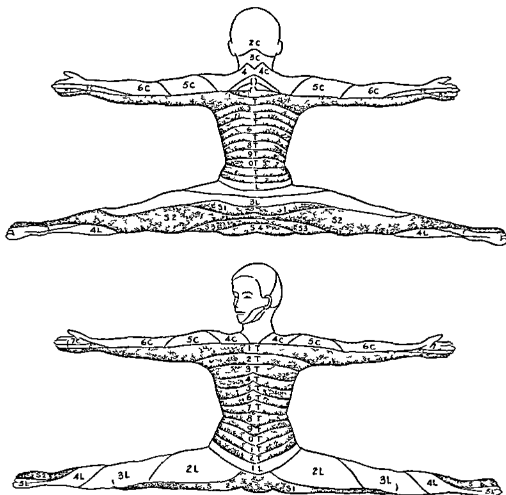


Fig 6 77 Cutaneous distribution of the spinal sensory nerves After Pottenger F M Clinical aspects of abdominal pain J A M A 102 341 (Feb 3 1934) Redrawn with permission of the author who credited Luciani s sketch from Bolk s data and the publisher

Neuritis may occur in any one of the spinal nerves The cutaneous distribution of the spinal nerves is shown in Figure 6 77 Muscular atrophies following injuries of the spinal nerves may be present

Spinal block if it results from tumor frequently causes an increase in the concentration of globulin in the cerebrospinal fluid below the tumor The fluid may also be xanthochromic The shade of yellow may vary, and occasionally the fluid above a block is decidedly yellow The cell count is usually normal but pleocytosis may occur if the tumor is situated in the spinal canal below the conus medullaris This may help in distinguishing neoplasms from inflammatory lesions

The presence of partial or total subarachnoid block is not pathognomonic of intraspinal tumor since previous attacks of meningitis acute myelitis injuries to the vertebrae or spinal deformities are all capable of interfering with the free flow of cerebrospinal fluid

Roentgenograms are made of anteroposterior and lateral aspects of the vertebral column supplemented by stereoscopic and oblique views localized at the level where a tumor has been suspected In general roentgenologic evidence of changes resulting from tumors of the spinal cord consists in shadows indicative of erosion of the vertebrae secondary to direct pressure invasion by the tumor destruction caused by benign or malignant tumor of the bone metastatic diseases or hyperostosis

In addition to the roentgenologic evidence of tumors which is apparent in routine examination of the spinal column roentgenoscopic and roentgenographic study by the use of radiopaque oil has furnished much additional information in diagnosis and localization of intraspinal tumors However radiopaque oil should be used only when tumors are suspected and the oil should be removed whenever possible

The manifestations of metastatic lesions may be the same as those of any benign tumor of the spinal cord except that the progress of a malignant growth is more rapid than that of a benign tumor Metastatic growths from carcinomas of the breast or the prostate gland are most frequent It must not be forgotten that a primary carcinoma of the breast may have been removed many years before metastasis gives evidence of its presence

DIAGNOSIS If there is demonstrated a level which limits distally the occurrence of both sensory and motor disturbances a single tumor of the spinal cord is probably present This lesion may be primary in the cord or secondary to a malignant lesion elsewhere in the body In the presence of such findings there is a history compatible with that of tumor of the spinal cord Special roentgenologic studies should be made by those who are skilled in such procedures and the roentgenograms should be interpreted by one skilled in such interpretations

When benign or malignant tumors can be removed completely the outlook is good provided the operation is performed early and the cord is not traumatized *Generally however the outlook is unfavorable despite the treatment*

DISEASES OF THE PERIPHERAL NERVES

Lesions of the peripheral nerve trunks are common causes of locomotor disability *Each peripheral nerve trunk has motor and sensory components* When the nerve is sectioned there is a complete loss of both of these functions Partial or progressive lesions which involve these component functions will vary

There are distinctions to be made in the diseases of the peripheral nerves the plexuses of nerves and the nerve roots When disease affects the nerve or the nerve trunk it is designated as neuritis the plexus as plexitis and the nerve roots as radiculitis

Disease of the nerve trunks of the spinal nerves occurs as mononeuritis and polyneuritis The mononeuritis is said to be an interstitial neuritis whereas the polyneuritis is caused by parenchymatous changes However several nerves may be affected by mononeuritis The causes for mononeuritis and plexitis occurring in one or several nerves are *infections such as leprosy syphilis trauma serum sickness* periarteritis nodosa diabetes and tumors of nerves such as neurofibromatosis (Recklinghausen s disease)

Injury of the Brachial Plexus The common type of brachial plexus injury is Erb s palsy which results from strong traction on the shoulder in the delivery of the head in breech delivery or from drawing of the head and neck away from the shoulders in the effort to deliver the shoulders in cephalic presentations The paraly sis is present immediately after birth

There are adduction and internal rotation at the shoulder extension at the elbow and often pronation of the forearm and some degree of flexion of the wrist When the arm is passively abducted it falls limply The flexors of the elbow and external rotators of the shoulder are inactive The grasping reflex is intact In mild injury there is no weakness at the wrist The biceps and radial reflexes are abolished As a rule it is impossible to demonstrate any loss of sensibility If recovery is delayed contractures soon develop in the antagonists of the paralytic muscles unless pre vented The head of the humerus becomes flattened and the margin of the acromion process becomes depressed in such a way as to prevent complete abduction

The *distal type* of brachial plexus palsy is less common It involves the intrinsic muscles of the hand with weakness of the long flexors of the wrist and fingers The grasp reflex is absent There is edema of the hand and there are trophic changes in the nails Loss of sensibility may be demonstrated in the hand The pupil is smaller than the opposite one and the homolateral eyelid droops although it can be opened The iris may remain unpigmented (Fig 6 79)

Complete paralysis of the entire arm and complete loss of sensibility almost up to the shoulder may be present There is partial arrest of development of the affected arm which is most evident in the hand (Fig 6 78)

Various bone injuries may be associated with brachial plexus palsies such as a fracture of the neck of the scapula fracture of the clavicle of the shaft of the humerus or separation of the epiphysis when they are due to postnatal trauma

Supernumerary cervical ribs may cause abnormal relationships in the neck and lower roots of the brachial plexus However supernumerary cervical ribs usually do not give rise to symptoms The symptoms of root compression may be due to pressure by a first rib when no supernumerary ribs are present Cervical ribs may be found in several members of a family and have indeed been traced through a number of generations (Macklin)

The *symptoms* of root compression rarely arise before the age of 20 years Despite the fact that the ribs are almost always bilateral the symptoms are almost invariably unilateral Drooping of the shoulders from poor muscular tone paralysis of the trapezius or wasting illness may precipitate the symptoms Women are affected three times as often as men The right arm is more likely to be involved than the left

On *examination* there is weakness of the hand and forearm with atrophy of the intrinsic muscles of the hand and of the superficial flexors of the wrist and fingers Paresthesias numbness and loss of cutaneous sensibility are found over the ulnar half of the hand and inner margin of the forearm The pain and thermal sensibility are reduced without demonstrable loss of tactile sensibility

The radial pulse may be small or absent In order to test for the presence of circulatory disturbances in a patient who has a cervical rib or ribs Allen Barker and Hines recommend that the patient be seated with the arms extended hori zontally with the elbows flexed until the forearms are perpendicular to the arms The head is turned as far as possible away from the side being examined A positive result of the test is indicated by diminution or disappearance of the radial pulse

Occasionally the subclavian artery may be spontaneously occluded resulting in gangrene of the fingers Manifestations resembling those present in syringomyelia or in Raynaud s disease are not uncommonly present in the scalenus anticus syn drome associated with cervical ribs

Lesions of Peripheral Nerves Small areas of analgesia surrounded by a larger area of tactile anesthesia suggest lesions of the *peripheral nerves* The size and location of the area indicate the nerve or nerves affected Traumatic injuries either laceration or pressure palsy are painless Interstitial neuritis is accompanied by severe pain and by tenderness of the nerve trunks Incomplete lesions of the peripheral nerves cause such syndromes as *causalgia* Symmetrically distributed tenderness of the calf muscles and nerve trunks and cutaneous anesthesia or hyper esthesia over the distal parts which fade out gradually over the more proximal parts are significant of polyneuritis

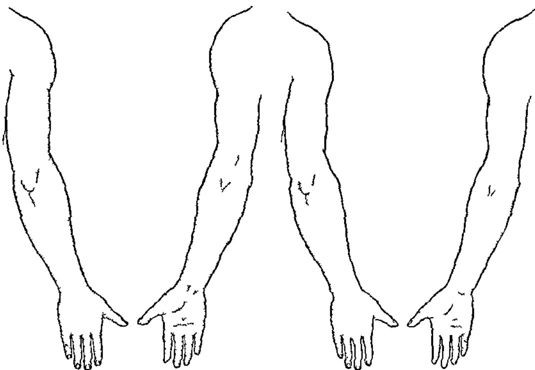


Fig 6 78 Area of sensory changes in lesions of the brachial plexus (C C_6 C_7 C_8 T_1)

Fig 6 79 Area of sensory changes when the lower part of the brachial plexus is affected (C_8 T_1)

Sensory changes in an area of the skin known to receive its nerve supply from a given nerve corroborate the finding of a weakened or paralyzed muscle or muscles, innervated by this same nerve In the diagrams the bordering areas are not depicted The limits of disturbances of light touch are included in the shaded areas which merge into the areas where sharp or dull cannot be differentiated

The Brachial Plexus The brachial plexus a great nerve plexus of the neck and axilla is formed by the union of the anterior branches of the lower four cervical nerves and the first thoracic nerve It supplies the whole upper extremity and its chief branches are the posterior thoracic suprascapular subscapular internal cutaneous musculocutaneous musculospiral circumflex median and ulnar nerves

The muscles supplied by branches of the brachial plexus and methods of testing have been given (pp 201 and 202) (Nerve supply, C_5 C_6 C_7 C_8 and T_1)

Sensory Changes in the Skin in Lesions of the Brachial Plexus and Peripheral Nerves of the Arm In Figure 6 78 are depicted the areas of sensory changes in the skin *in complete lesions of the brachial plexus* The level of segmental integration of the brachial plexus is C_5 C_6 C_7 C_8 and T_1

The sensory area of skin disturbance *when only the lower roots* C_8 T_1 of the brachial plexus are affected is shown in Figure 6 79

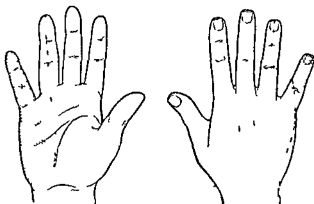


Fig 6 83 Area of sensory changes of the hand Ulnar nerve distal to origin of its dorsal branch

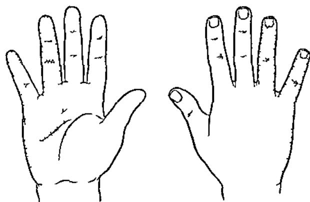


Fig 6 84 Maximal area of sensory changes in skin of the hand *observed in lesions proximal to the* origin of the dorsal branch of the ulnar nerve

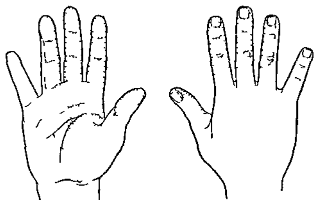


Fig 6 85 Maximal area of sensory changes of hand observed in lesions of the median nerve

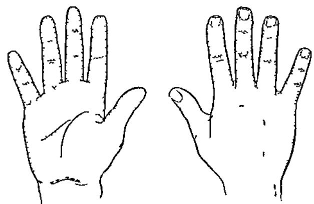


Fig 6 86 Sensory changes in hand when both median and ulnar nerves are affected

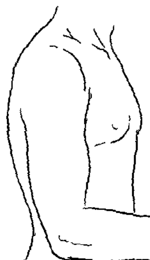


Fig 6 80 Area of sensory changes which attend injuries of the axillary nerve (C_5 C_6)

In *diagnosis* the roentgenographic demonstration of cervical ribs in association with slowly developing atrophy of the muscles of the hand pain and other disturbances of sensation and pulse volume are *confirmative*

Sensory Changes in the Skin Resulting From Lesions of the Peripheral Nerves of the Arm and Hand The area of distribution of sensory change when the *circumflex (axillary) nerve* is injured is revealed in Figure 6 80 If the lesion involves the axillary nerve too there will be paralysis of the deltoid muscle

The *radial nerve* may be injured proximal to its branch the dorsal antibrachial cutaneous nerve When the nerve is injured above this nerve the approximate area of change in cutaneous sensibility is outlined in Figure 6 81

When the radial nerve is injured distal to its dorsal antibrachial cutaneous branch and the musculocutaneous nerve is injured too a combined lesion exists The sensory changes in the skin are diagrammed in Figure 6-82 The radial nerve also is the *motor nerve* to the brachioradialis (C and C_6) extensor carpi radialis supinator (C, C_6 and C) triceps (C_6 C) extensor digiti communis extensor digiti V proprius extensor carpi ulnaris abductor pollicis longus extensor longus and extensor proprius and extensor pollicis brevis muscles *(see Muscle Testing of the Arm and Hand pp 213 to 220)*



Fig 6 81 Area of sensory change in injury of the radial nerve proximal to the dorsal antibrachial cutaneous nerve

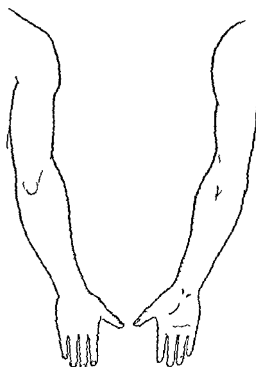


Fig 6 82 Area of sensory change in injury of the radial nerve distal to the dorsal antibrachial cutaneous nerve

The approximate area within which sensory changes may be present in disturbances of the *posterior cutaneous nerve of the thigh* is revealed in Figure 6 90

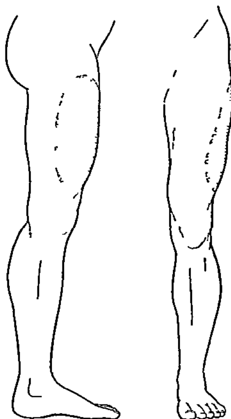


Fig 6-89 Areas of sensory changes in lesions of the lateral femoral cutaneous nerve (L_2, L_3)



Fig 6-90 Area of sensory changes in injuries of the posterior cutaneous nerve of the thigh (S_1 S S_3)

The approximate area within which sensory changes may be detected in lesions of the trunk of the *sciatic nerve* is revealed in Figure 6 91

The area within which sensory changes occur in lesions of both the *sciatic* and the *posterior femoral cutaneous nerve* is revealed in Figure 6-92

An area within which sensory changes may be revealed in lesions of the *lateral popliteal (peroneal) nerve* above the origin of its musculocutaneous branch is shown in Figure 6 93

An area within which sensory changes are present in lesions of the *deep peroneal nerve* is shown in Figure 6 94

Sensory changes in lesions of the *sural nerve* are within the area revealed in Figure 6 95

The area of sensory changes in lesions of the *medial popliteal or tibial nerves* is revealed in Figure 6-96

The cutaneous changes observed in the skin of the hand or injuries of the ulnar nerve depend on whether the injury is situated distal or proximal to the dorsal branch of the ulnar nerve When the lesion is situated distal to its dorsal branch, the skin changes are shown in Figure 6 83 When the lesion is situated proximal to its dorsal branch the maximal area of cutaneous changes over the hand ensues (Fig 6 84)

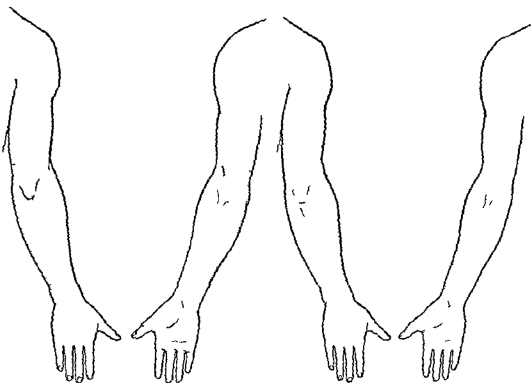


Fig 6 87 Area of sensory changes resulting from injuries of the medial antibrachial cutaneous nerve Origin brachial plexus

Fig 6 88 Area of sensory changes resulting from injuries of the lateral antibrachial cutaneous nerve

The ulnar nerve is the motor nerve to the flexor carpi ulnaris lumbricales III and IV flexor digitorum profundus adductor pollicis flexor digiti brevis V, opponens digiti V abductor digiti V and interossei muscles of the hand (see Muscle Testing of the Arm and Hand pp 213 to 220)

The *median nerve* supplies sensory fibers to the skin of the hand when maximal injury has been sustained by this nerve the areas of sensory changes are revealed in Figure 6 85

When both the median and the ulnar nerves are injured the area of sensory changes over the hand is revealed in Figure 6 86

The median nerve is the motor nerve to flexor carpi radialis pronator teres opponens pollicis lumbricales I and II flexor pollicis longus palmaris longus flexor digitorum sublimis flexor digitorum profundus pronator quadratus abductor pollicis brevis and flexor pollicis brevis muscles (see Muscle Testing of the Arm and Hand pp 213 to 220)

Injuries of the median cutaneous nerve of the forearm cause sensory changes in the skin as illu trated in Figure 6 87

Injuries to the lateral antibrachial cutaneous nerve of the forearm (a branch of the radial nerve) cause changes in the skin as revealed in Figure 6 88

Sensory Changes in the Skin Resulting From Lesions of the Peripheral Nerves of the Leg and Foot The area within which sensory changes may be found in lesions of the *lateral femoral cutaneous nerve* is revealed in Figure 6 89

Allergy Occasionally polyneuritis develops in association with serum sickness The muscles supplied by the fifth and sixth cervical roots are usually most severely affected

Physical Agents A large variety of poisons may cause polyneuritis Lead neuritis is perhaps the commonest form Arsenical neuritis was formerly common but has been rare since the medicinal use of arsenic has been discontinued Some of the sulfonamide derivatives may cause severe polyneuritis

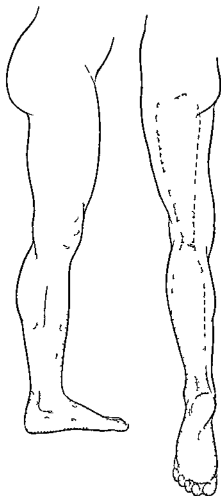


Fig 6 92 Area of sensory changes when the posterior femoral cutaneous nerve and the sciatic nerve are both affected

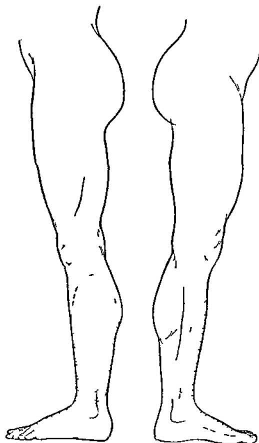


Fig 6 93 Areas of sensory changes in lesions of the common peroneal nerve

Diabetic neuritis is often clinically latent—that is to say the patient may not make any complaints but examination reveals tenderness of calf and plantar muscles and absence of the ankle jerks In more severe diabetic neuritis pains in the legs reminiscent of the lightning pains of tabes may be complained of the knee jerks may be lost and the degree of postural sensory loss may produce an ataxic gait and lead to an erroneous diagnosis of tabes dorsalis

Polyneuritis may be associated with acute *porphyrinuria* hemochromatosis and amyloidosis

It is thought possible that chronic alcoholism and the cachexia of malignant disease and of chronic tuberculosis may lead to defective absorption of the relevant vitamins This is not substantially proved

Lesions of Spinal Roots If the spinal roots are injured the sensory loss corresponds to the distribution of the posterior roots and a radicular anesthesia The area of tactile anesthesia is smaller than that of *relative analgesia and there is often* so much overlap of the tactile fibers that even in extensive root lesions anesthesia cannot be demonstrated For instance, in neuritis of the brachial plexus there is *usually no anesthesia* but there are usually much pain and tenderness and cutaneous

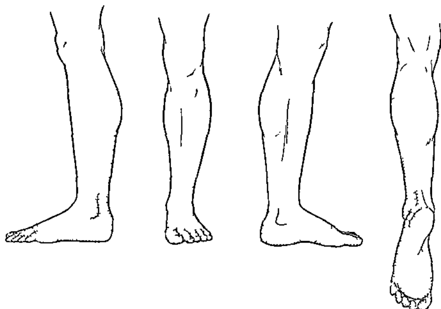


Fig 6 91 Area of sensory changes in lesions of the sciatic nerve (trunk) Origin L_4 L_5 S_1 S S_3

hyperesthesia Herpes zoster causes relative anesthesia over one or more dermatomes The pain may be relatively widespread but the anesthesia if present is confined to the general region of the cutaneous lesions The postherpetic neuralgia often causes much suffering in elderly persons See Figure 6 77 page 373

Radiculitis Radiculitis is defined as an inflammation of the root of a spinal nerve Usually the greatest part of the inflammation is situated between the spinal cord and the intervertebral canal The spinal cord is somewhat movable in the spinal canal so that when the spinal column is flexed the cord rises slightly and when it *is extended the cord sinks backward When there is meningitis or radiculitis* flexion of the neck or elevation of a forcefully extended leg causes pain The Lasegue Kernig and Brudzinski signs are based on the presence of radiculitis

In radiculitis as in neuritis irritative phenomena of the motor nerves occur There are fascicular twitchings which may end in atrophy of the muscles involved if the irritation is progressive or is long continued

Irritative phenomena of the sensory root are manifested by pain

Polyneuritis (Multiple Peripheral Neuritis) Multiple peripheral neuritis or *polyneuritis is a bilaterally symmetric affection of the peripheral* nerve trunks to the limbs which involves both motor and sensory fibers In some instances cranial nerves may be affected

Infections Infections are the commonest cause of polyneuritis *In childhood* diphtheria is responsible for a large percentage of all cases It is usually recognized by the associated paralysis of the pharynx and soft palate and the loss of accommodation In the more acute and severe infections the *Landry syndrome of paralysis occurs Acrodynia* may also be associated with polyneuritis

SYMPTOMS The various etiologic forms of polyneuritis do not differ essentially in their symptomatology The variation comes in the rapidity of development of the original disease and of recovery from it Special symptoms which have been described as being peculiar to an etiologic variety of polyneuritis are symptoms of the original disease

The symptoms common to all forms of polyneuritis are the manifestations of a paralysis of lower motor neuron type The legs are affected earlier and more severely than the arms The muscles below the knee and distal to the elbow are more severely affected than the proximal muscles There is drop foot and in severe instances wristdrop The muscles are tender to pressure In long standing polyneuritis wasting and contractures and deformities ensue

EXAMINATION There are diminution and later loss of tendon reflexes All forms of sensibility are impaired varying in degree and like the paralysis maximal over the distal extremities The trunk is rarely involved Early in the severely acute polyneuritis there is tachycardia In some instances there is cardiac dilatation and even fatal syncope

DIAGNOSIS The diagnosis is made from the findings on examination If possible the diagnosis should indicate the etiologic agent

Acute Febrile Polyneuritis (Landry s Paralysis) Acute febrile polyneuritis differs in no essential from what is described as Landry s paralysis The term Landry s paralysis indicates an acute spreading and often fatal paralysis characterized by the absence of severe or constant changes in the nervous system

Landry s paralysis and acute febrile polyneuritis commences by a brief period of febrile disturbance with headache malaise and sleeplessness Often gastrointestinal disorders may precede the appearance of the paralysis The paralysis which develops rapidly is of a lower motor neuron type flaccid with loss of tendon jerks Commonly there is paresthesia in toes and feet accompanying the development of paralysis which spreads up the legs and often involves the arms The abdominal and back muscles and the facial muscles may be affected The muscles of respiration are less severely weakened than the limb and facial muscles except in fatal illness

A pulse rate increase above 110 to 120 is a serious prognostic omen

If the patient does not die the paralysis begins to diminish clearing first in the muscles least severely weakened A general debility and exhaustion ensue The tendon reflexes slowly reappear

Interstitial Neuritis or Perineuritis Interstitial neuritis is a local inflammatory reaction involving chiefly the interstitial connective tissue of the nerve trunk which is distinguishable from the generalized degenerative process of polyneuritis

There are interstitial neuritides which are associated directly with active foci of infection and others which are not so associated An interstitial neuritis may be directly associated with arthritis abscesses lymphadenitis nasal sinusitis otitis cellulitis erysipelas tonsillitis infected teeth and septic ulcers In many instances when the nerve lies in proximity to the site of infection the inflammation reaches it by direct lymphatic extension

Debilitating infections such as pneumonia typhoid fever or influenza may be followed by interstitial neuritis

The mechanism of production of an interstitial neuritis without direct association with foci of infection is not clear

The process commences by inflammation and swelling of the nerve which then becomes dense as the result of proliferation of connective tissue The axis cylinders are not severely affected

The symptoms begin with deep and superficial pain which follows the course of the nerve trunk and is projected into the region of cutaneous distribution The pain is sometimes constant but more often intermittent and paroxysmal The disease is extremely chronic

On examination it is observed that movements and postures which serve to place

Nutritional Disorders It is well known that vitamin deficiency more especially de ficiency of vitamin B will cause polyneuritis Walshe believes that Beriberi is probably induced by a toxic product formed in the course of the disordered carbohydrate metab olism that ensues when carbohydrates are consumed in large quantities without the

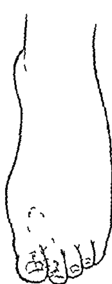


Fig 6 94



Fig 6 95

Fig 6 94 Area of sensory changes in lesions of the deep peroneal nerve

Fig 6 95 Area of sensory change in lesions of the sural nerve

normally occurring natural vitamins with them Some as yet unknown component of the vitamin B complex is essential to the carbohydrate metabolism of the nerve cell Despite the clinical evidence at hand there are yet important gaps in the chain of evidence linking B avitaminosis with the development of polyneuritis

The role of ethyl alcohol in the etiology of polyneuritis may be associated with lack of vitamin B

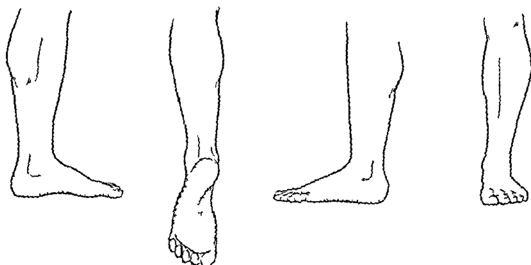


Fig 6 96 Area of sensory changes in lesions of the tibial nerve

Constantly disorders of funct on in instances of acute polyneuritis precede struc tural changes since there may be complete or relative absence of visible changes in the nervous system in *certain rapidly fatal instances of multiple neuritis* such as beriberi and Landry s paralysis In long standing fatal multiple neuritis degenerative changes of wallerian type are found in the axis cylinders of the affected nerves The nerve cells and fibers of the cerebral cortex may also show a degree of degenerative change The heart shows infiltration with fat and myocardial degeneration

and is partially anesthetic The nerve trunk which supplies the region of the lesion begins to become enlarged and palpable Various signs of neuritis slowly develop

EXAMINATION On examination the usual findings in leprosy comprise some one or many of the following The fifth nerve frequently is affected There may be patches of anesthesia over the forehead the bridge of the nose and the cheeks which are associated with regions of depigmented skin and macular nodular (tuberculoid) or ulcerating nodular cutaneous lesions

The *maculae* of leprosy involve the corium of the skin particularly the hair follicles and the sweat glands The bacillus of leprosy is difficult to demonstrate in these lesions The *tuberculoid lesions* of leprosy occur as discrete ulcers separated by normal tissue As the disease advances these nodules may become continuous masses pushing other structures aside as they expand With the expansion of the lesions the skin is thinned and hyperkeratinization with scaling and hypopigmentation or white spots in the skin develops

At this stage of the disease the patient loses first the ability to discriminate between heat and cold next sensibility to pain and finally tactile sense

Frequently the anesthesia is bilateral and sometimes it is symmetric Neuroparalytic keratitis may ensue All modalities of sensation may be affected but with a certain amount of dissociation The degree of dissociation of cutaneous sensation observed in syringomyelia is not present There is usually palpable thickening of the superficial nerve trunks especially of the supra-orbital nerve Localized palsies of the facial muscles are common and characteristic The ulnar nerve and the common peroneal nerve are affected early in the course of the disease Atrophy and weakness appear in the intrinsic muscles of the hands and feet and slowly give rise to various types of flexion deformity including the so-called Simian hand The ankle jerk and finger reflexes are lost and the knee and biceps reflexes are preserved The plantar response is abolished and the abdominal reflexes are preserved

Disorders of vasomotor and secretory function and trophic disturbances are constant The hands and feet are cold and blue Sweat secretion is lost and the hair often falls out Dry necrosis of the fingers may be present but it is commoner to find concentric atrophy of the small bones of the fingers and toes so that the digit grows smaller and shorter Neuropathic joints are sometimes present The ends of the digits may slough away and leave only stumps of the fingers

The nasal mucous membranes are frequently affected often with early perforation of the nasal septum The mucous membranes of the tongue pharynx and larynx may be involved Involvement of the conjunctiva cornea and iris which leads to blindness is frequent The liver and spleen frequently are enlarged and the secondary deposit of amyloid or amyloidosis is exceedingly common in the late stages of this disease Amyloid involving the kidneys gives the symptoms of nephritis

DIAGNOSIS A definite diagnosis of leprosy must be based on finding of the bacilli in the nasal secretions or in the skin lesions Material aspirated from affected lymph nodes will often reveal the bacilli For diagnostic purposes a person who has thickening of the skin around the forehead eyes nose and lips accompanied by other cutaneous lesions enlargement of lymph nodes in the inguinal and axillary regions and regions of anesthesia over the extremities or regions of anesthesia with rather severe pain in the arms or legs has leprosy

The differential diagnosis of leprosy involves the following conditions cutaneous syphilis nasal or oral leishmaniasis yaws and Boeck s sarcoid Recklinghausen s disease (the form described as forme fruste) and syringomyelia

Nodular leprosy is likely to run a more rapid course than neuritic leprosy and to lead to death within 10 years Neuritic leprosy usually lasts longer Spontaneous remissions are described

Alcoholic Neuritis The first symptoms of alcoholic neuritis are cramps and pains in the calf and plantar muscles at night There may be aching and fatigue in

the affected nerve under tension increase the pain The nerve trunk is tender and the tenderness is most severe over the inflamed segments Sometimes the nerve trunk is palpable In severe and prolonged instances of the disease there are paralysis and atrophy of the muscles loss of reflexes and anesthesia

A diagnosis of interstitial neuritis or perineuritis requires history and evidence of a severe or debilitating infection or of neuritis if the tenderness is confined to the nerve trunk and if stretching the nerve causes pain Roentgenographic examinations are of value in excluding diseases of the bones and joints A conservative prognosis is always given

Leprosy Leprosy is due to infection by an acid fast bacillus *Bacillus leprae* (*Mycobacterium leprae*) which morphologically resembles the tubercle bacillus

Regularly in the neural or anesthetic type of leprosy the peripheral nerve trunks have nodular thickenings as a result of proliferation of connective tissue and cellular infiltration The bacilli are sometimes present in large numbers in the innermost lamellae of the perineurium and in the endoneural septa The number of bacilli is independent of the degree of tissue reaction In some cases the reaction is relatively acute with active inflammatory reaction and edema but as a rule the process is chronic and the thickening of the nerve is due chiefly to fibrosis

Degeneration within the posterior roots and of the exogenous fibers of the posterior columns occurs relatively often The posterior root ganglia may be the seat of degeneration and of fibrosis Nonspecific degenerative changes in the brain have been described

In the cutaneous form of leprosy there are firm intradermal and subcutaneous nodules over the face and other exposed surfaces Ulceration and secondary infections are common Mutilation of the parts is due mainly to trauma from lack of sensation and to ulceration secondarily infected In all forms of leprosy lesions usually develop in all organs except the voluntary muscles The cartilage of the nose may be destroyed The mucous membranes of the mouth nose and eyes are badly injured The lymph nodes may enlarge and become granulomatous The testes are often destroyed Nephritis is common

DISTRIBUTION Leprosy probably arose in Central Africa It is common in the Balkans Asia Asia Minor India China Siberia Africa Iceland Japan Australia and the Philippines In the United States small numbers of cases are found in the Gulf region along the Pacific coast and in the Great Lakes region

Leprosy is common among children whose parents have the disease If the children are removed from their parents immediately after birth the incidence of the disease is low Congenital leprosy is rare Infection is thought to be acquired through the skin or through the nasal mucosa and to reach the skin by way of the blood stream The incubation period of leprosy is unknown

Two varieties of leprosy the nodular form and the maculo anesthetic or neuritic form are described However mixed types are common

SYMPTOMS The early symptoms of leprosy may be the same irrespective of the form of the disease which is about to develop Irregular fever sweating weakness and epistaxis may be present or a local cutaneous lesion may appear without other symptoms

In the nodular form erythematous lesions develop on the face forearms thighs and buttocks which become indurated nodules and eventually ulcerate However there appear on the skin some spots which are often hyperesthetic at the onset but eventually become anesthetic Nodules appear on the mucosa of the upper part of the respiratory tract and in the conjunctiva and cornea The ulcerations progress and great mutilations appear as the result of trauma and secondary infection and extend These mutilations are the basis for the classic descriptions of the disease

In the maculo anesthetic form the cutaneous lesions develop singly rather than in successive outbreaks In these the macular lesions are white or yellowish and attain 1 to 3 inches (about 2 5 to 8 cm) in diameter These depigmented spots appear on the limbs trunk or face They often have an erythematous border Often there is neuritic pain at the onset The skin affected by the macules does not sweat

voluntary control after complete palsy is accompanied by an increased muscle tone After the nerve has regenerated an asymmetry of the face may remain This asym metry is not evidence of incomplete regeneration but evidence that nerve fibers have been misdirected and there thus remains a disturbance of function

The diagnosis of facial paralysis (Bell s palsy) is usually certain However it is well to be aware of the fact that if the palsy has developed slowly an intracranial neoplasm especially pontine glioma is likely

The prognosis is based on the severity of the lesion Rarely will regeneration fail and the facial muscles atrophy When regeneration fails there is extreme relaxa tion of the tissues and an unsightly deformity ensues

Guillain Barre Syndrome *(Polyneuritis With Facial Diplegia) The cause of* the Guillain Barre syndrome is unknown An infective agent is not proved

There are no specific pathologic observations Polyneuritis and degeneration of nerve roots may be found with occasional changes in the cord

The syndrome is characterized by symmetric flaccid paralysis with abolition of the tendon reflexes and with subjective sensory symptoms without objective change The patients are afebrile but may have had an antecedent febrile illness All show a symmetric flaccid paralysis involving particularly the proximal muscles of the limbs and extending in severe forms to the cranial nerves with a predilection for the seventh nerve Facial paralysis exists but involvement of the other facial nerves is unusual Increase in the protein content of the cerebrospinal fluid occurs frequently but is not a pathognomonic sign Differentiation from poliomyelitis is important and not difficult

Tumors of Peripheral Nerves ***Neurofibromatosis of Recklinghausen*** The disseminated neurofibromatosis of Recklinghausen is characterized by multiple tumors arising from the sheaths of the spinal or cranial nerves by cutaneous pig mentation and tumor formation by abnormalities in the bones and frequently by evidences of defective development and other malformations

The disease recurs in successive generations of affected families and therefore is be lieved to be the expression of an abnormal germ plasm The tumors of the nerve trunks are true connective tissue growths When there are intracranial neurofibromas there may be meningeal growths usually termed dural endotheliomas Gliomas and areas of gliosis often occur in association with neurofibromatosis of the generalized type In a large percentage of all cases of glioma of the optic chiasm there is associated Recklinghausen s disease At the age of puberty the tumors become somewhat more numerous and larger than they have been previously The cutaneous lesions are usually symptomless

Examination reveals irregular areas of yellow or brown pigmentation on the skin Interspersed in the areas between these large spots are small circular brown-ish areas resembling freckles The skin is often dark Numerous cutaneous tumors (molluscum fibrosum) of all sizes occur ranging from 1 mm in diameter to tumors measuring 2 to 3 cm in size These cutaneous tumors are pigmented soft or firm and a few are pedunculated

Neurofibromas occur on both the cranial and the spinal nerve roots In some instances they seem to be confined to the two eighth nerves In others many tumors are found in the cranium The vagus nerve is most frequently affected and the fifth nerve stands next in frequency At times the brain stem and cerebellum are com pressed by large neurofibromas

Tumors occurring on the spinal nerve roots give rise to signs of compression of the spinal cord or cauda equina They cannot be recognized as neurofibroma before operation even though cutaneous tumors are present It is unusual to find intra cranial growths in the presence of pronounced cutaneous lesions

Plexiform neuromas extending diffusely through the tissues may occur over the cranium causing elephantiasis of the scalp Plexiform neuroma may occur in the eyelid orbit sella in the bones of the skull and in the arm or leg

those muscles during activity Tingling and numbness in the feet and toes are often present These symptoms may or may not be associated with other indications of chronic alcoholism for instance looseness of the bowels morning nausea loss of appetite tremor of the hands and rapidity of the pulse and flushed cheeks Gradually weakness of the legs and subjective sensory symptoms become more prominent and an altered mental state (polyneuritic psychosis or Korsakoff s psychosis) ensues The memory is so bad that the names even of familiar objects are forgotten there may be disorientation as to place and time The patient may indulge freely in fic titious reminiscences

Examination in the initial stage reveals marked tenderness to pressure of the plantar and calf muscles The knee jerks may still be present but the ankle jerks are absent There is cutaneous hyperesthesia a moderate degree of sensory impairment (pain touch temperature vibration) and postural modes of sensibility over the feet and toes Sensory loss spreads up the limbs and impairs postural sensibility so that the ambulant patient is likely to be ataxic and to sway when in the Romberg position

In the advanced stages of the disease there is great loss of power in the distal segments of the limbs Often there is drop foot manifested by a high stepping gait but less often wristdrop Paralysis progresses and complete invalidism may ensue The muscles shorten and contractures and deformities develop The cutaneous hyperesthesia may make all handling of the limbs painful and difficult

Arsenical neuritis very closely resembles alcoholic neuritis even in the occasional presence of the polyneuritic psychosis This disease is exceedingly rare since the decrease in the employment of arsenical therapy

Neuritis of the Facial Nerve (Bell s Palsy) Bell s palsy follows an infectious interstitial neuritis or it results from exposure to cold There is a hemorrhagic inter stitial neuritis involving that part of the nerve which lies in the temporal bone in close relation to the middle ear

In Bell s palsy resulting from interstitial neuritis there is pain localized usually in the ear extending into the cheek or into the occiput In Bell s palsy resulting from chilling the palsy precedes the pain

Variations in symptomatology depend on the site of the lesion If the lesion is distal to the point at which the chorda tympani separates from the seventh nerve taste is not affected Involvement of the geniculate ganglion is accompanied by a herpetic eruption within the concha and external auditory canal If the lesion is situated between the geniculate ganglion and the brain stem there is no overflow of tears and the eye remains dry The presence of any one of these symptoms forces the assumption to be made that the lesion is probably situated inside the cranium and is not a simple Bell s palsy

On examination there is a hypotonicity of some and paralysis of other muscles of the face on the side of the paralyzed nerve The forehead and the cheek are smooth The corner of the mouth sags The forehead cannot be wrinkled the eye closed or the corner of the mouth retracted The patient cannot whistle Labial enunciation is affected There is always an accumulation of food outside of the teeth on the affected side Taste is lost over the anterior portions of the tongue The lower lid falls away from the bulbar conjunctiva and tears run down the cheek for the lacrimal punctum is no longer in position to collect them Blinking of the eyelid is associated with twitching of the mouth

When the palsy is complete the affected side of the face sags and the lip and cheek often become edematous As a rule the pain ceases within a few days

Within a few weeks examination reveals some return of muscle tone and later there is power of movement The recovery is prompt and complete if the palsy is partial If paralysis has been complete return of power is slow The return of

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A benign neurofibroma rarely undergoes sarcomatous degeneration and metastasis

The presence of cutaneous lesions or multiple subcutaneous nodules offers as a rule no difficulty in diagnosis since their nature is evident In the central type of the disease the diagnosis cannot be made before operation Roentgenograms of the bones may be helpful in diagnosis

There is a *forme fruste of Recklinghausen s disease* characterized by the pigmented skin lesions without tumors

It is unusual to see a patient who is seriously disabled by this condition Intracranial or intraspinal tumors are serious since they are almost always so numerous that they cannot be removed

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7

DISEASES OF THE ANORECTAL REGIONS AND GENITALIA

THE ANORECTAL REGIONS

THE PELVIC OUTLET

The topographic markings of the structures which limit the area within which the anus rectum and the external genitalia are situated are known as the pelvic outlet The pelvic outlet is bounded anteriorly by the pubis and laterally by the descending rami of the pubis and the ascending rami of the ischia Posteriorly is the coccyx and near its lateral limits are the spines of the ischia The greater and the lesser sacrosciatic ligaments form a diamond shaped space which is divided into two triangles The anterior one is the urogenital and the posterior one is the anal triangle (Fig 7 1) These triangles may be visualized by passing an imaginary line

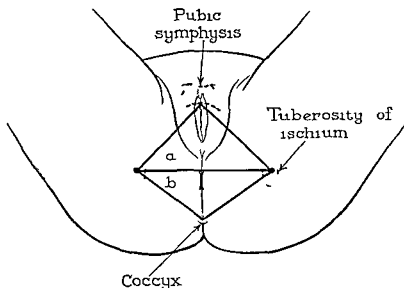


Fig 7 1 Pelvic outlet a urogenital triangle b anal triangle

from the anterior portion of one ischial tuberosity to the opposite one Such a line passes about ½ inch (1 27 cm) anterior to the anus This imaginary line passes through the dorsal portion of the central point of the perineum At the central point of the perineum the anteroposterior and transverse muscles meet This point is sometimes designated the perineal body

THE PERINEUM

The Perineal Fascia and Muscles It is clinically important to remember that there are several layers of perineal fascia and muscles which may have implications in perineal infections and injuries The first layer of the superficial fascia is continuous with the

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on the perineum anterior or lateral to the anal verge and the cyst often does not contain hair

Unless complicated by infection the condition is symptomless although at times there may be slight discharge Infection in due time is the rule The symptoms then are those of superficial pain due to abscess swelling tenderness and fever In many cases there is a history of numerous acute episodes and surgical operations Often relief follows simple incision

A skin lined orifice or orifices oval or round situated in the midline over the sacrococcygeal region are usually indicative of pilonidal cyst or pilonidal disease Such openings often but not always are hair-containing In pilonidal cyst or sinus a probe passed through the orifice passes upward whereas in a fistula in ano the probe will pass downward These sinuses and cysts tend to recur even after careful attempts have been made to remove them surgically

Hidradenitis Suppurativa Hidradenitis suppurativa is a chronic inflammatory disease of the skin and subcutaneous tissue in those regions in which the apocrine sweat glands are situated namely the axillary mammary inguinal genital and perianal regions This condition may resemble according to Jackman pilonidal disease

The diagnosis of perianal manifestations of hidradenitis suppurativa is usually not difficult if the examiner thinks of this disease when he encounters what appears *to be extensive anal fistula or pilonidal disease with many sinuses Examination* with the patient under anesthesia may be necessary to distinguish pilonidal cysts or anal fistula from hidradenitis suppurativa

Dermoids and Teratomas Dermoids and teratomas are often midline tumors attached to the periosteum of the sacrum and lying between the sacrum and the rectum The presence of the tumor often is not suspected until the size of the mass gives symptoms of pressure or unless the tumor is discovered on routine examina *tion The tumor may burst into the rectum* and the patient *may give a history* of expelling a tuft of hair and teeth Occasionally a tooth or a tuft of hair may be visualized on protoscopic examination The diagnosis can be established by biopsy if teeth or hair is not present

THE ANUS AND RECTUM

The anal canal extends from the rectum to the anus or its opening on the skin a distance of 1 to 1½ inches (2 5 to 3 8 cm) *It begins at the level of the levator ani* muscles and ends at the apex of the prostate gland which is directly in front of it and with the tip of the coccyx behind and a little above With the body vertical the anal canal has its axis inclining upward and forward toward the bladder as soon as the sphincter ani is passed the axis of the rectum changes to upward and backward toward the hollow of the sacrum In introducing a speculum it should always be inclined first anteriorly and then posteriorly Opposite the level of the levator ani the circular muscular fibers increase to form the internal sphincter This extends down the anal canal for a distance of approximately 1 inch (2 5 cm) and ends above the skin margin The external sphincter surrounds the lower part of the canal and stretches in a spindle shape from the tip of the coccyx to the central point or tendon of the perineum Anteriorly it blends with the fibers of the levator ani and the other muscles of the perineum It is a thick powerful voluntary muscle and extends outward to the mucocutaneous junction

Mucous Membrane The upper half of the mucous membrane of the anal canal has six or eight longitudinal ridges or folds (*columns of Morgagni*) Between the lower ends of these columns are small hollows called the crypts (of Morgagni) and the free edges of the mucous membrane guarding the crypts are the anal valves

The sacculation of the rectum is produced by three creases or crescentic folds called the rectal valves or valves of Houston Of these the middle is the largest It springs from the right anterior quadrant about 2 to 2¼ inches (5 1 to 5 7 cm) above the margin of the anal canal The superior and inferior valves spring from the left posterior quadrant a short distance above and below the middle valve At the juncture of the rectum and

surrounding parts and the dartos muscle The second layer of the *superficial fascia* (Colles fascia) a fibrous underlayer is continuous with the scrotum and passes forward to form the subcutaneous (Buck s) fascia of the penis covers the spermatic cord and is continuous with the deep layer of the superficial fascia of the abdomen (Scarpa s fascia) The anterior layer of the triangular ligament blends with the posterior layer of the deep *superficial fascia* *The deep layer of the triangular fascia* is a continuation downward of the pelvic fascia the obturator fascia and passes onto the levator ani muscles and closes the gap between these muscles

The superficial perineal space is situated between the crura of the penis covered by the ischiocavernosus muscles

The deep perineal space lies between the layers of the triangular ligament and contains the compressor urethrae muscle embedded in which is Cowper s gland with its duct which empties into the bulbous urethra

Extravasations of urine or blood in moderate amounts occur from injuries to the *penis anterior to the triangular ligament and thus pass into the superficial space and* are confined there Since this space extends forward and is continuous over the scrotum the extravasating fluid passes forward and distends the scrotum In greater extravasations the urine may extend to the penis and along the spermatic cord over the crest of the pubis to the surface of the anterior abdominal wall and laterally to the flanks but not down the thighs

When rupture of the membranous urethra occurs urine or blood may pass between the layers of the triangular ligament and be present in the superficial tissues anterior to this ligament In more serious rupture urine escapes into the deep perineal space and thence back between the prostate and rectum or thence anteriorly behind the symphysis pubis into the perivesical space between the peritoneum and transversalis fascia

The Ischiorectal Fossa The ischiorectal fossa is situated between the tuberosity of the ischium and the anus Its main affections are from abscesses from the rectum It has an anterior process which runs forward between the prostate gland internally and the ischiopubic ramus externally The posterior process is its deepest extension and communicates from side to side between the coccygeal attachment of the external sphincter muscle of the anus The ischiorectal fossa is enclosed by the levator ani and coccygeus muscles on the inward side and by the obturator internus muscle on the outward side

Development of the Organs Incident to the Formation of the Perineum The bladder is first recognizable at a stage when it receives on each side a mesonephritic duct and a ureter Soon the four ducts namely the two ducts of the mesonephron and the two ureters become separated and the ureters move laterally The bladder when it begins to form is tubular in shape then it expands and is saccular in shape with the elongate urachus at its apex The urachus is continuous with the umbilicus and may in rare instances maintain this connection between the umbilicus and the bladder throughout life

The female urethra is formed by an elongation of the original connection between the primitive bladder and the urogenital sinus The male urethra is a counterpart of the female organ and in addition the pelvic portion of the urogenital sinus is added to form the prostatic and membranous urethra The phallic portion of the urethra adds the cavernous urethra

The prostate gland originates from multiple outgrowths of the urethral epithelium The outgrowths are soon surrounded by connective tissue and smooth muscle fibers The Skene glands in the female are homologous to the prostate but are few in number and situated near the external urethral outlet

The bulbourethral glands (Cowper) of the cavernous urethra of the male are homologous to the vestibular glands (Bartholin) of the female

The seminal vesicles arise as outpouches from the mesonephron and gain muscular walls from the mesenchyme There is no homologous organ in the female

Pilonidal Cyst and Sinus Outside of the pelvic outlet in the midline of the coccygeal region if there has been a failure of union of the ectodermic layer of the embryo there occurs below the surface an inclusion of hair growing tissue termed pilonidal cyst However as N D Smith has pointed out this cyst may be situated

ectoderm is on or near the surface of the body Surface structures have as an important function the protection of the organism from the adversities of the environment and at least a part of this protection is obtained through the ability to perceive pain Structures arising from the mesoderm and entoderm when stimulated do not give rise to definite localized pain A sharp pain in anorectal disease points to a pathologic condition involving the anus

Imperforation and Associated Malformations Imperforation and the malformations associated with imperforation constitute most of the congenital disorders of the rectum The various types of imperforations may be enumerated as the imperforations of the rectum with vesical posterior urethral uterine or vaginal outlet In an occasional instance there may be a spinal outlet In females the genital outlets may open into the rectum Also the rectum may be imperforate for lack of an anal outlet or the rectum may have difficulty in emptying owing to stenosis of the anus and thus may empty through the vagina

IMPERFORATE RECTUM The rectum may be absent or its descent may be arrested at any point distal to the promontory of the sacrum The usual point of arrest is at the peritoneal reflection A fibrous cord may extend from the apex of the rectum to the anal cul de sac or from the imperforate rectum to the bladder or the urethra

The first symptom of an imperforate rectum after birth is absence of bowel motions This is followed by the symptoms of obstruction of the colon

On examination there may be a normal appearing anus and anal canal or the anus may be absent If the anal canal is present the well lubricated little finger when inserted into the rectum reveals a shallow rectum completely occluded

Imperforate rectum with vesical outlet is a rare condition The communicating passage is narrow and lined with mucosa It opens into the bladder often near the trigone The diagnostic symptom is fecal (meconium stained) urine On examination the finger is stopped at a shallow depth It is then obvious that the source of the feces stained urine is an opening of the rectum into the urinary system Communications other than from the rectum to the bladder are usually in the posterior urethra

In *imperforate rectum with posterior urethral outlet* the passage from the imperforate rectum may open into the prostatic or membranous urethra Some of the patients who have this malformation attain adult life The diagnosis of imperforate rectum with vesical outlet or urethral outlet is definitely established by endoscopic examination

In *imperforate rectum with vaginal or uterine outlet* the communication usually opens into the vestibule of the vagina The symptoms and signs are the same as those of an imperforate rectum with the addition of the passage of feces from the vagina Often these openings may be adequate for drainage of the rectum and consequently the patients may reach adult life The diagnosis is made by digital or endoscopic examinations once the condition is suspected

Imperforate rectum with spinal outlet is an obstetric curiosity These babies do not usually survive

In those who have a *urogenital outlet in the rectum* the vagina may be normally formed but opens into the rectum and in some cases the abnormality is unnoticed until the menstrual flow is established The bladder is absent and the ureteral orifices may be at the peritoneal reflection in the rectum Urine in the rectum is usually well tolerated without an ascending urinary infection The diagnosis is made by endoscopic examination of the rectum once the condition is suspected

IMPERFORATE ANAL CANAL When there is an arrest or irregular development of the proctodeum anomalies of the anus result The anus may be completely absent and the median raphe may extend in an unbroken line to the coccyx In the absence of the anus however the site for it usually is marked by a slight depression a corrugated cutaneous prominence or a discoloration surrounded by rugae In still other instances the anus may be partially or fully formed with obstruction in any part of it Anomalies of the anus may be limited to this stricture or may occur in association with those of the rectum

the sigmoid flexure there is another fold on the anterior wall which tends to obstruct the view when examinations are made These valves are composed of connective tissue and circular muscular fibers covered with mucous membrane

The combined length of anal and rectal canal is 4 to 5 inches (10 2 to 12 7 cm)

RECTAL EXAMINATION The finger can palpate the anal canal and rectum for a distance of 4 inches (10 2 cm) from the surface Anteriorly in men as soon as the finger passes the sphincters the apex of the prostate can be felt also the membranous urethra particularly if it contains a bougie or sound The prostate can be outlined and its size determined If the prostate is not enlarged the base of the bladder above can be palpated and the tip of the finger will reach the rectovesical pouch From the upper or posterior edge of the prostate and extending from near the midline upward and outward are the seminal vesicles Just to the outer side of the upper end of the seminal vesicles are the lower ends of the ureters Posteriorly the coccyx and the hollow of the sacrum can be felt Laterally the finger can explore the region of the spine of the ischium the sacrosciatic foramina and the tuberosities

Ataxic Sphincter This is a condition in which the external sphincter lacks tone owing to degenerative changes in the posterior nerve roots and posterior columns of the spinal cord often as a result of tabes dorsalis or of syringomyelia The anus appears normal on inspection but when it is separated it remains partly open instead of closing instantly as it does normally and then very slowly resumes its usual state of contraction When tabes dorsalis is not otherwise suspected this symptom often gives the first indication of the disease

In some patients an ataxic sphincter in the absence of any demonstrable lesion of the peripheral nerves or the central nervous system is due to a prolonged use of laxatives

Rectal Shelf Masses Properly the term rectal shelf has been employed to designate that condition in which a palpable mass impinging on or attached to the anterior rectal wall causes a ledge or shelf to be felt Such a mass deserves serious evaluation because it may call attention to a lesion remote from the rectum The mass may resemble a primary carcinoma of the rectum and thus lead to an error in diagnosis and prognosis A patient may consult a physician because of symptoms produced by the rectal shelf or a pararectal mass without relating these symptoms to any which result from the primary lesion or the reverse situation may be encountered The commonest causes of pararectal masses among women are lesions of the genital system and among men lesions of the gastrointestinal tract and the stomach (carcinoma) in particular

Congenital Malformations of the Anus and Rectum ***Persistence of Cloacal Duct*** In persistence of the cloacal duct there is a narrow outlet from the rectum to the raphe of the perineum scrotum underside of penis prostatic urethra or bladder In the female the opening may be along the perineal raphe or into the vestibule of the vagina

The rectum develops from the entoderm and the mesoderm the anus the proctodeum develops as an invagination of the ectoderm The primitive rectum and the primitive anus gradually approach each other Absorption of the epithelial layers of the proctodeum and the primitive rectum permits the joining of the lumina

The conjunction of the rectum and the anus at a point in front of the posterior end of the gut leaves a cul de sac This cul de sac and the connecting canal are largely absorbed during fetal life Imperfect absorption is the cause of a congenital posterior rectocele The region where this posterior cul de sac and the communicating canal are situated is a favorite site for the occurrence of dermoid cysts and other teratoid tumors of the rectococcygeal space

As has been indicated the anus and the rectum developmentally and functionally are distinct organs arising from different layers of the embryo and thus are subject to diseases inherent to the tissues from which they arise The diseases of each organ present symptoms which are unlike those of diseases of the other This is particularly true of the symptom of pain The anus arises from ectoderm Any structure which arises from

hemorrhoids and polyps Some large papillae may be drawn out of the anus by the palpating finger Attempts to pull these away with the examining fingers cause acute pain and hemorrhage

Abscesses and Ulcers ***Anorectal Abscesses*** The diseases such as fissures ulcers abscesses fistulas and abrasions are often initiated by breaks in the continuity of the anorectal lining membranes resulting from trauma and not by direct invasions of the tissue by bacteria viruses or parasites

An anorectal or a rectal abscess develops from an injury made by the passage of formed feces of large caliber often containing hard pellets which tear one or more of the anal crypts These tears are common and usually heal promptly Occasionally such a tear does not heal promptly and then an inflammation of the crypt occurs spreads to the surrounding tissues and results first in the condition known as cryptitis The cryptitis either heals or proceeds to abscess formation

The extent of the inflammation and the structures affected determine the type of anorectal abscess present For instance if the pus forms above the levator ani muscle the abscess is either a *superior pelvirectal abscess* or a *retrorectal abscess* depending on which of these spaces is occupied An abscess posterior to the levator ani muscle is an *ischiorectal abscess*

The superior pelvirectal and retrorectal inflammations originate outside the rectum These inflammations are frequently extensions of an infection of the adjacent pelvic urogenital organs or of the intestines Occasionally an ileitis may originate such an abscess

The studies by Hill and co workers of structure of the anal glands offer a possible explanation of the spread of infection through these glands There is a wide individual variation as to number depth of penetration and contour of the anal glands Lack of complete canalization of the terminal portion of the anal glands was observed in 3 newborn infants In some of the specimens from adults the cells lining the anal glands presented apical cytoplasmic vacuoles which stained positive for mucin

Various existing traumatic bacterial and constitutional factors unite to permit infection to spread by way of the anal glandular system The extension of anal glands upward beyond the anorectal line into the rectal submucosa offers an explanation of the occasional occurrence of abscess and fistula in this region The deep penetration of an anal gland extending into the internal sphincter points to possible avenues of invasion along muscle bundles fascial planes and adjacent structures to these lines of cleavage A definite glandular cyst in an adult supports the claim that abscess and fistula may develop without communication with the anal canal

SYMPTOMS In the superior pelvirectal and retrorectal abscesses pain is deep but not intense Prostration is often profound for large quantities of pus can accumulate in these spaces There may be intermittent fever These abscesses are often the cause of the so called pyrexia of unknown origin In a few who have these abscesses the temperature may be as high as 105 F (40 5 C) or more

If the abscess has had its origin in a pelvic organ for instance an ulcerative colitis or from an ileitis or if it has originated in the prostate and extended the whole side of the buttocks may become exceedingly tender reddened and swollen There often are fever and an accompanying diarrhea

An abscess arising in the anus which extends to the perianal tissues is characterized by local redness swelling pain and hot skin similar to the manifestations of a superficial abscess elsewhere

EXAMINATION On examination the anal orifice and buttocks often appear normal On digital examination of the rectum above the anorectal line a soft tender mass may be palpated The leukocytes vary in number from normal to 30 000 per cubic millimeter of blood

The anus may be completely formed but occluded at the anorectal line by a thin elastic diaphragm of fibrous or mucocutaneous tissue due to the persistence of the anorectal membrane In these infants there are no bowel motions Examination often reveals an impulse or a fluctuation of the distended rectum

The diagnosis of imperforate anus is usually made by inspection and palpation

Congenital Stenosis of Anus The normal anus of an infant should admit without difficulty the lubricated little finger The canal may be narrow throughout or segmentally The narrowing may be so slight that the passage of soft feces causes no difficulty but harder masses give rise to constipation and straining

The diagnosis is made by digital examination Normally the anal canal varies in caliber and due allowances are given for these normal variations

Anal Ulcers (Fissures) Anal fissures and ulcers are usually manifested in the third fourth and fifth decades Occasionally they are present in children but rarely *in the aged Two men have anal fissures or ulcers to each woman so affected* Nine of every 10 anal ulcers or fissures occur in the posterior portion of the canal

Anal fissures or ulcers are almost always caused by fecal trauma However trauma also may result from sharp particulate matter or a fishbone in the feces A common source of anal trauma is the introduction of a syringe tip An uncommon source but nevertheless one that does occur is sodomy Sodomy may be followed *by gonorrheal proctitis and ulcers Injury and destruction of an anal crypt may result* in ulceration

The characteristic symptoms are bleeding with bowel motions and pain occurring from a few minutes to a half hour after bowel movement The pain continues for variable lengths of time and then entirely or partially subsides until the next bowel movement There is a wide variation in the degree of pain Some patients may have little or no pain from an anal ulcer or fissure

On digital examination the anus is exceedingly tender When the sphincter is spread open the ulcer is seen to be oval or circular and just within the grasp of the muscle Often it has a superficial fistulous tract leading toward the skin surface and the teatlike process known as the sentinel pile at the skin margin of the ulcer

A chancre of the anus may closely resemble the usual type of anal ulcer Anal chancres occur more frequently in women than in men

Cryptitis The anal crypts are small pockets distributed about the anorectal line Inflammation of these pockets is termed cryptitis This inflammation has been designated infected hemorrhoids

The anal crypts have their openings directed inward They are subjected to trauma by the passing feces Following trauma mucosal abrasions within the crypts may originate an inflammatory reaction These reactions often terminate in pararectal abscess

The symptoms of cryptitis closely resemble those of ulcer or fissure There is pain commencing after a bowel movement Pains referred to the perineum the urogenital region the sacrum and coccyx or down the legs are common On examination there is tenderness along the anorectal line

Cryptitis is not usually an inflammation of a single crypt but an inflammatory reaction that has originated in a crypt and then spread along the anorectal line The proctoscopic findings are characteristic enough for diagnosis by an experienced examiner

Papillitis Papillitis is an inflammation of the anal papillae The anal papillae are often found to be *hypertrophied This enlargement without symptoms* is considered to be an incidental condition

Enlarged papillae may be torn by the stool resulting in sharp pain in the anus and bleeding Usually there are no symptoms of papillitis

On rectal palpation the finger may perceive soft or firm teatlike projections at the anorectal line Large papillae may cause some examiners to suspect internal

is differentiated from perianal tuberculosis by biopsy Usually however the diag nostician has little doubt of the differentiation even without laboratory aid

The prognosis is never good The condition usually is secondary to tuberculosis in some other part of the body and even when the ulcers heal there is a constant danger of reinfection Often too the disease advances despite all treatment

Tuberculosis of the Rectum Tuberculosis of the rectum practically always is secondary to an infection elsewhere in the body the lungs being the usual primary seat The rectum is involved more rarely than the intestinal regions above but more frequently than the anus

Three forms of rectal tuberculosis are described the miliary the ulcerative and the hyperplastic

Miliary tuberculosis of the rectum is rare and always is part of a generalized tuberculosis Multiple tubercles form beneath the mucosa and these coalesce and slough to form irregular ulcers Sometimes these ulcers will burrow beneath the mucosa to form small fistulas

In *tuberculous ulceration* of the rectum the tubercle bacilli are carried to the rectal mucosa through the lymphatics and there form tubercles As these break down and coalesce they form small ulcers The edges of these ulcers undergo a progressive degeneration which leads to a constant increase in their size until there are large ulcerations

Secondary infection of the ulcers may cause them to perforate the musculature of the rectum and even of the peritoneum resulting in tuberculous abscesses of the perirectal tissues and subsequent fistulas which may extend into the bladder or other contiguous organs with the production of a tuberculous peritonitis

The symptoms of tuberculous ulceration of the rectum are cramplike pains during bowel movements often accompanied by tenesmus and frequent stools sometimes numbering as many as 30 during the 24 hours Contained with the stool is a mucopurulent blood tinged fetid discharge The discharge scanty in the be ginning increases in quantity until it becomes profuse When the discharge is copious it is thin and irritating to the anal skin

On examination the ulcers are irregular gray and rough appearing with slightly thickened irregular and undermined edges The mucosa between the ulcers is likely to be inflamed and edematous

The diagnosis of the disease is based on identification of the tubercle bacilli by cultural means guinea pig inoculation and on the characteristic appearance of the ulcers and the fact that the disease occurs in a patient who has active pulmonary tuberculosis or tuberculosis of some other part of the body

Hyperplastic tuberculosis of the rectum is a rare form of the disease It is char acterized by a localizing thickening of the intestinal wall due to a round cell infiltra tion and the massive deposit of fibrous tissue This condition commonly known as tuberculoma may resemble malignant disease The treatment is surgical

Tuberculous Pararectal Abscess The tuberculous pararectal abscess has an insidious onset When it has fully developed there is no pain or tenderness over the infected region Usually the only symptom will be distortion of the part from the accumulation of pus The condition always is secondary to tuberculosis in some other part of the body usually the lungs and it may occur during the course of a tuberculous ulceration of the rectum An accumulation of fluid beneath the skin of the buttocks and near the anal margin without acute symptoms arouses suspicion of a tuberculous abscess particularly in a person who has pulmonary tuberculosis

Tuberculous Fistulas These fistulas result from the evacuation of pus from a tuberculous abscess The fistulas may connect with the skin of the buttocks or with any organ adjacent to the rectum and anus

Syphilis There may be itching early but the chancre or ulcer is painless unless it is within the grasp of the sphincter or is complicated by a mixed infection

When the abscess has extended to involve the perianal tissues particularly in the presence of an ileitis or chronic ulcerative colitis there may be swelling and redness of the skin and induration of the tissues of one side of the buttocks When these manifestations are present rectal examinations may be impossible because of the tenderness and pain

The superficial abscess can be recognized from the mildness of its manifestations Rectal examinations are painful but usually can be performed if care is exercised during the examination In the presence of the superficial abscess the inside of the rectum feels normal and it is not tender

DIAGNOSIS The diagnosis of an acute superficial abscess is made from the presence of pain tenderness and redness of the affected parts Fluctuation may be present

The diagnosis of deep abscess such as pelvirectal and retrorectal abscesses may be uncertain The prostration high fever and leukocytosis and often the sensation of weight and heaviness in the pelvis draw attention to this region These symptoms accompanied by the presence of a tender rectal mass justify a presumptive diagnosis In due time after exercise of judgment and caution an exploratory incision may be required for both diagnosis and proper treatment

Anorectal Tuberculosis A tuberculous infection of the rectum and adjacent tissues occurs in a patient who has swallowed infected milk or sputum and thus an opportunity has been rendered the bacillus to invade the tissues of the rectum The tubercle bacillus may invade the anorectal regions from an anorectal focus through the lymphatics through the blood stream and by direct extension from contiguous organs from inside the pelvis

Tuberculosis of the Anus *Tuberculosis of the perianal skin* occurs as lesions miliary ulcerative lupoid and papillary (verrucous) The lupoid and papillary forms are rarely observed

Miliary lesions of the anus are a part of an advanced and generalized infection in some other part of the body Multiple minute nodules form beneath the epidermis and gradually coalesce and break down to become the ulcerative type of tuberculosis Diagnosis is established by biopsy and guinea pig inoculation The prognosis is poor

Ulcerative tuberculosis of the anus is the commonest form of the disease It may be secondary to a focus in the lungs or intestines but is more likely to be a primary infection than is tuberculosis of the rectum Small nodules form in the skin and then coalesce and disintegrate to become ulcerations The ulcerations may be single or multiple and at times are epitomized by massive ulceration that involves the entire perianal region The individual ulcers usually are superficial but may involve all of the layers of the skin Their shape is irregular and they have a distinct margin depressed below the surface of the raised base Sloughing and undermined edges are characteristic

Patients who have severe exudative and destructive pulmonary disease are more subject to anal fistulas than patients who have the productive type of the disease The perianal abscess either precedes or concurs with a progressive or regressive modification of the pulmonary process Patients who have severe tuberculosis may exhibit multiple anal fistulas surrounded by granulation tissue extensive zones of infiltration and detachment of skin at the external openings whereas in the fibrous type of pulmonary tuberculosis the fistulas are benign When ulcerative pulmonary tuberculosis exists specimens obtained by scraping the fistulous tract often yield tubercle bacilli In the fibrous type of pulmonary tuberculosis with negative sputum positive results are less frequent Tissues surrounding anal fistula often yield positive evidence of tuberculosis The anatomic and clinical observations indicate that the perianal tuberculous abscess is probably of hematogenous origin

The diagnosis is established by biopsy and guinea pig inoculation Epithelioma

opening but no external opening Many variations of these fistulas exist (Fig
Often there are multiple external openings which communicate with each othe
with an internal opening

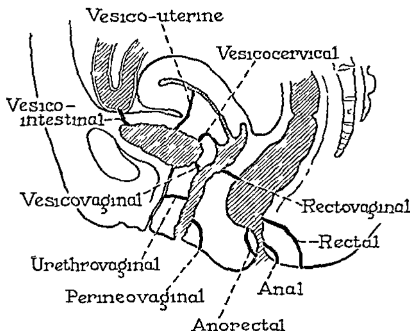


Fig 7 2 Common sites for anal anorectal rectovaginal vesicocervical vesicou perineovaginal and urethrovaginal fistulas

The genesis of fistula is a pyogenic organism with the formation of an absces organism present is usually the staphylococcus not the tubercle bacillus

Inflammation of the crypts of Morgagni extending to the perirectal tissue terminate in formation of abscess The abscess may rupture into a pelvic org through the skin of the buttocks and thus establish a fistula An abscess of this typ rupture into the vagina the uterus the urethra the prostate gland or the bladde *external opening of these fistulas* does not bear a constant relationship to the p of the internal opening There may be many external openings over both buttock thighs

Symptoms Fistulization about the anus and rectum is characteristicall ondary to the rupture of an acute abscess somewhere about the rectum genitalia The symptoms of the abscess are obtained from the history and are summarized by the presence of pain redness swelling and fever The abscess o spontaneously or was lanced and relief from symptoms followed the dra After the rupture or lancing of the abscess a small opening persisted in the bu from which there was constant oozing of a purulent or serous discharge wh time ceased After a lapse of time another acute abscess developed This seq of events may be repeated with the maintenance of a single fistula or with f tion of each new abscess new fistulas may be added until the entire buttock perirectal tissues are involved in fistulas

Examination On examination the external opening of the fistula is r found Often a probe may be gently worked through the fistulous tract u emerges through the internal opening in the rectum under direct visuali through an endoscope Often however the tract is tortuous and it is impossi work the probe through the tract in an unanesthetized or on occasion an ane ized patient The probable internal opening may be discovered by palpating the finger from within the rectum

If the lesion is single and fairly characteristic of chancre a presumptive diagnosis can be made A definite diagnosis depends on positive results of darkfield examination Serologic reactions of the blood are seldom positive before the second week or later

Condyloma Latum This is a secondary manifestation of syphilis occurring within the anus or in the perianal skin and characterized by wartlike elevations occurring singly or in groups

A pearly white flat oval or rounded patch begins on the skin surface and multiplies These plaques have a tendency to form large fungating masses which may encircle the anus and extend to the vulva or to the scrotum The masses are broad flat warty elevations with sharply defined edges which rise at right angles from the skin The surface is flat and covered with a glistening grayish necrotic membrane Diagnosis depends on a positive Wassermann reaction combined with the history of a primary lesion and the presence of the wartlike plaques just described

Rectal Injuries and Foreign Bodies ***Rectal Injuries*** Spontaneous rupture of the rectum may occur as the result of crushing blows to the abdomen or pelvis and from accidental dilatation from the nozzle of a pneumatic hose entering the rectum Many different sorts of anorectal injuries are sustained in farm accidents for instance falling astride a fence post and falling on the handle of a pitchfork

The symptoms are severe abdominal pain prostration shock and hemorrhage Diagnosis is not difficult as a rule and should be made early so that surgical intervention may be performed immediately to prevent or to alleviate peritonitis

Foreign Bodies Foreign bodies find lodgment in the rectum usually by being swallowed but they may be inserted through the anus In those who have mental diseases all manner of objects may be packed in the rectum Rarely they may gain entrance from a contiguous organ or they may form within the intestinal tract (gallstones fecaliths)

Symptoms are extremely variable and depend to a great extent on the size and shape of the object within the rectum Small objects such as fishbones which become caught within the anus cause a sudden sharp and constant pain

The diagnosis depends entirely on rectal examination Endoscopic and roentgenologic examination should be resorted to whenever rectal examination leaves the least doubt as to the nature extent or situation of the offending body

Anal Stenosis and Contracted Anus (Pectenosis and Anal Fibrosis) The inflammatory stricture follows extensive removal of the anal skin and superimposed infection may result from (1) hemorrhoidectomy (2) trauma (3) the injections in the treatment of pruritus and hemorrhoids (4) the excision of fistulas and anal condylomas (5) chronic ulcerative colitis (6) lymphopathia venereum and (7) irradiation therapy The stricture often is tubular and may be from 1 inch to 3 or 4 inches (2 5 cm to 7 6 or 10 2 cm) in length and the strictured lumen may be measured in millimeters

Bowel movements are difficult and painful In some cases blood and pus are passed from the rectum with stool or sometimes as a leakage independently of bowel movement

The anus and rectum are of small caliber and very tender so that digital examination may be impossible Often a proctologist can pass an instrument of small caliber for endoscopic examination of a patient who cannot permit a digital examination

The diagnosis is evident from the foregoing findings The treatment is dilatation except in those who have chronic ulcerative colitis active lymphopathia venereum or irradiation therapy

Anorectal Fistula A *complete anorectal fistula* implies an internal opening in the anus or rectum or at the anorectal line and an external opening in the skin surface near the anal orifice or in the buttocks An *incomplete fistula* has an internal

On inspection and palpation there is a livid firm tender swelling beneath the smooth shiny skin After the acute symptoms have subsided the swelling will seem shotlike a characteristic that will persist for a long period There are sometimes erosion of the overlying skin and bleeding from the thrombotic hemorrhoids A swelling on one side of the anus which is tender and painful firm and livid beneath smooth shiny skin is diagnostic

Internal Hemorrhoids The etiologic factors capable of producing internal hemorrhoids are heavy lifting standing posture sedentary habits heredity portal hypertension constipation straining at stool and infection A low grade long standing infection weakens the venules and veins of the rectum so that the weakened lining of the veins ruptures and acute thrombosis results After repeated thromboses occur there is prolapse which is constant and after bowel movements the prolapsing mass is replaced regularly with attendant trauma The prolapsing mass increases in size and protrudes through the sphincter muscle The muscle may lose its tone owing to continued pressure and long standing disease

Without thrombosis internal hemorrhoids are painless Bleeding occurs at bowel movements Blood may squirt in a small stream as the patient strains The quantity of blood lost at any bowel motion is seldom great but the staining of the water in the bowl may give the impression of a large hemorrhage Internal hemorrhoids will occasionally bleed spontaneously for instance while the patient is pursuing the daily tasks

A prolapse of the mucous membranes may occur at bowel motion The prolapsing mass often retracts without assistance When the prolapse is marked however the mass will not return within the lumen of the rectum and must be replaced by the patient A prolapse hemorrhoid which remains outside the anal canal will cause a constant dragging pain in the rectum and there may be perianal edema

The examining finger can feel large internal piles or sclerosed piles but not small ones The diagnosis is made by proctosigmoidoscopic examination

Prolapsed Internal Hemorrhoids Prolapsed internal hemorrhoids may get caught in the grasp of the anal sphincter and become edematous To replace prolapsed hemorrhoids ask the patient to lie on the left side Bathe the hemorrhoids with cold water to reduce the edema Apply petroleum jelly liberally with cotton Exert pressure on the protrusion with one hand while the other hand is easing the constriction of the edematous folds of the anus If the edema is so great that the hemorrhoids cannot be returned it may be necessary to anesthetize the sphincter with some local anesthetic agent

If the hemorrhoidal mass cannot be thus replaced the condition is termed strangulated piles or hemorrhoids This condition requires hospitalization and appropriate treatment

Proctitis and Proctosigmoiditis Proctitis is an inflammation of the rectum Sigmoiditis is an inflammation of the sigmoid flexure that portion of the colon which extends from the crest of the ilium to the level of the third sacral vertebra Inflammation of both the rectum and sigmoid is known as proctosigmoiditis

Proctitis or proctosigmoiditis may result from constitutional disorders such as diarrhea and fever caused by the exanthems influenza and hepatic diseases Any chronic disease accompanied by diarrhea may cause proctosigmoiditis All of the acute infectious dysenterial diseases are accompanied by proctosigmoiditis

The manifestations are often those of hemorrhoids the hemorrhoidal irritation having been produced by tenesmus and the frequent passage of watery bowel motions which may contain blood and mucus

Diagnosis is established by the presence on endoscopic examination of a reddened inflamed mucous membrane of the rectum and rectosigmoid Time and repeated observations may be necessary to be sure that the disorder is not an early stage of chronic ulcerative colitis

DIAGNOSIS The diagnosis is made from the findings revealed by physical and endoscopic examinations

Hemorrhoids Hemorrhoids appear externally and internally (Fig 7 3) External hemorrhoids are derived from the inferior hemorrhoidal vein and are situated below the anorectal line Internal hemorrhoids are derived from the superior hemor

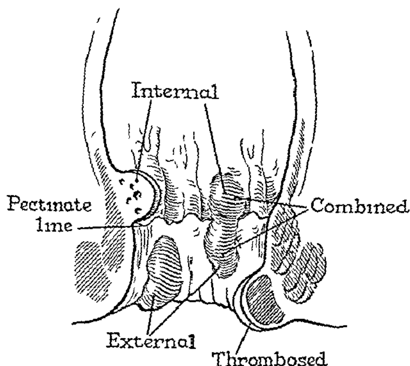


Fig 7 3 Hemorrhoids

rhoidal vein and are situated above the anorectal line They are covered with mucosa

External Hemorrhoids External hemorrhoids comprise skin tabs and varicosities of the anal veins Since man walks on his hind legs the cause of external hemorrhoids is postural They are not present in four footed animals The incidence of external hemorrhoids is high in both men and women

Skin tabs are objectionable because of the difficulty they cause in efficient cleansing of the parts They usually do not produce symptoms

Varicosities are seen as bluish swellings just within the anal canal or at the anal margin and are soft and compressible

The foregoing described diagnostic findings are unmistakable and symptoms are usually absent

THROMBOTIC EXTERNAL HEMORRHOIDS Thrombosis of external hemorrhoids results from a strain or traumatism to the thin walled anal veins Coughing sneezing lifting or straining at stool may cause one or more of the coats of a diseased vein to give way to be followed by formation of clot within the walls of the vessel or in the surrounding connective tissue Once the thrombus is formed it may remain and the swelling and pain gradually cease in from 3 to 10 days If rapid distention of the veins takes place rupture may follow

Immediately following the rupture there is a feeling of continuous discomfort Sometimes throbbing burning pain in the anus ensues and continues As the pain and soreness develop it becomes more and more difficult to sit in comfort or to move the bowels Pressure on the tumor is painful The discomfort continues to be severe for 2 or 3 days and then gradually subsides

but will be found to consist of more proximal segments of the bowel These masses are smooth soft and feel like the contents of a hernial sac

Pruritus Ani Anal itching is a symptom most frequently of nervous tension and irritability The unusual causes of pruritus ani are diabetes infestation with pin worms (see Diseases of the Colon Chapter 13) pediculosis pubis fungous or other infections of the skin general dermatosis and infections of the anorectal region A severe anal pruritus may result from the application of salve for the treatment of hemorrhoids

Localized itching may be mild and present only occasionally or it may be continuous and severe The itch is more intense after the patient has retired for the night or perhaps after he or she has been sleeping for half of the night The itch may be agonizing and there is an overwhelming desire to scratch the affected area The patient scratches himself unconsciously when awake or during light sleep

Unbearable and constant itching in time because of the consequent loss of sleep and persistent irritability will lead to physical fatigue and mental exhaustion

On examination interspersed about the anus will be excoriations from scratch ing The skin about the anus is often reddened swollen and either dry or moist In advanced severe pruritus ani the skin assumes a leathery appearance Lichenification a pale and blanched appearance because of a loss of pigmentation ensues The radiating skin folds are likely to be edematous and hypertrophied and the sulci between to become cracked or fissured

The diagnosis of pruritus ani depends on the history of itching about the anus and the findings revealed on physical examination

Tumors of the Anorectal Regions ***Benign Tumor*** *Verruca acuminatum of the anus* has the synonymous names anal wart papilloma and verruca Warts and papillomas that occur about the margin of the anus and within the anal canal are often called venereal warts Condyloma latum however is the only one of venereal origin

The papillomas consist of connective tissue and blood vessels covered with stratified epithelium They arise from hypertrophy of the papillae of the skin and are caused by irritating discharges such as occur in gonorrhea hemorrhoids proc titis and colitis Verruca acuminatum is an excrescence which begins on one side of the anus and often spreads rapidly until it surrounds the anus as a large cauliflower like tumor attached to the skin by many pedicles The growths are soft and pliable of a pale pink color and bleed freely when injured They are distinguished from epitheliomas by the absence of induration at the base or ulceration of the surface

Verruca acuminatum and papillomas are to be distinguished from the highly malignant papillomatous forms of anal epitheliomas which are situated at the anal *verge or within the anal canal Likewise they are differentiated from protruding* hypertrophied anal papillae which may protrude from the anus The diagnosis is established by biopsy and histologic study

Malignant Tumors ANAL CARCINOMA (Epithelioma) Anal carcinoma arises from the squamous cell epithelium of the skin and mucocutaneous junction and thus is an epidermoid carcinoma having all the characteristics of carcinoma of the skin and of lesions of mucocutaneous junctions such as the lip or vulva Most of the growths are ulcerating lesions but they may appear as cauliflower like masses pro truding from the anus

The complaint may be of anal discomfort with or without relation to stool and the passage of large quantities of fresh blood In addition the patient may report the presence of a tag or growth within or near the anal orifice which has suddenly en larged and become painful These features are highly suggestive of an anal lesion and the odds are greatly in favor of anal fissure since anal epithelioma occurs only rarely Nevertheless the possibility of epithelioma must be considered in all middle aged patients of either sex

Gonorrhea Gonorrhea of the rectum is uncommon The gonococcus may gain entrance to the anus and rectum by inoculation of pus from an infected urethra by pederasty or by the contamination of instruments Diagnosis is made only on the basis of positive cultures

Prolapse Protrusion and Procidentia of the Rectum and Sigmoid Colon Prolapse or procidentia is a descent of one portion of the rectum or anal canal into or through another In complete prolapse there is an intussusception of the rectum

Prolapse of the rectum is classified as *incomplete or partial prolapse* when only the mucous membrane is involved and *complete prolapse or procidentia* when all of the coats of the bowel form a part of the prolapse When the colon becomes involved the prolapse is termed a *colonic procidentia* since the anus does not have three coats A patient can have an anal prolapse but not an anal procidentia

The common causes of prolapse and procidentia are either intrinsic or extrinsic to the bowel The intrinsic conditions causing prolapse and procidentia are hemorrhoids polyps neoplasms foreign bodies and sodomy Extrinsic conditions are parturition tumors and displacements of the uterus hypertrophy and tumors of the prostate visceroptosis intra abdominal tumors prolonged vomiting coughing or crying tightly applied belts and corsets certain violent exercises urethral calculus and urethral stricture and phimosis

Prolapse occurs in children less than 5 years of age and in the aged who have debilitating disease and nutritional disorders with a loss of ischiorectal fat

Disease of the spinal cord resulting in paralysis or relaxation of the anal sphincter or levator ani muscle such as tabes dorsalis predisposes to prolapse and procidentia

In the beginning there is protrusion only at stool a prolapse At first the protrusion is of slight degree The protrusion may gradually become greater until several inches or more of the bowel may remain outside Subsequently the external sphincter becomes more and more atonic until it becomes difficult or impossible to keep the bowel in its proper place Often the condition is intermittent Discomfort *described as a bearing down sensation is present whenever the mass* is outside of the body and it may be present at other times

The mucosal prolapse is a redundancy of the mucosa which protrudes through the anus at stool It is bright red moist and glistening and has small hemorrhagic areas on its surface Protrusion through the anus occurs at stool and the protruded mass may return within the sphincter spontaneously or may remain outside until pushed within Protrusions which have been present for a long period may *remain* outside of the anus When this occurs the reddened congested mucosa often becomes eroded and ulcerates With the passage of time the mucosa turns pale and assumes a leathery appearance and the sphincter loses its tone

DIAGNOSIS Prolapse of the rectum which does not extend beyond the anus is diagnosed by proctoscopic examination An abnormal looseness or wrinkled swelling of the mucosa will be seen and this may involve a segment of the rectum or its entire circumference

Procidentia recti the displacement of the rectum with all of its coats into its lumen or through the anus is distinguishable by the presence of the musculature shown by concentric rings in the mass These rings constitute the differential point *from a prolapse protruding* from the anus in which folds radiating from the anus are seen in the mass The mucosa of the procidentia after long exposure outside of the body becomes thickened and firm

A procidentia of the rectum which does not protrude from the body can be palpated with the finger and is felt as a mass with a lateral sulcus filling the rectum

A sigmoidal procidentia presents the same characteristics as those of the rectum

The disease is manifested by a cluster of small erosions (eroded vesicles) on the glans penis or corona sometimes associated with a group of typical herpetic vesicles on the shaft Herpetic infection of the genitalia of women as of men may be primary or recurrent The primary disease is manifested by the appearance of superficial erosions on the labia majora labia minora and distal vaginal mucous membrane These ulcers are tender and covered by a grayish yellow membrane The annoying discomfort usually subsides within a few days but may persist for a week or two There is a tendency for the disease to recur with similar erosions of clustered vesicles revealing an ulcerated region covered by the grayish yellow membrane

Phimosis Phimosis classified as (1) congenital and (2) inflammatory is ordinarily a symptomless defect in the preputial orifice characterized by muzzling or closure of the skin over the distal end of the penis so that the skin cannot be drawn back over the glans Phimosis late in life may give rise to symptoms as follows (1) retention of smegma (2) preputial concretions (3) adhesions (4) balanoposthitis (5) paraphimosis (6) gangrenous belanoposthitis and (7) carcinoma

Smegma accumulates behind the adhesions slowly in some persons rapidly in others The retention of smegma is a factor which predisposes to balanoposthitis and probably carcinoma Retention and ammoniacal decomposition of drops of urine mingled with smegma are the usual cause of the formation of preputial concretions and odors The tight foreskin as the result of low grade infection forms adhesions to the corona and often over much of the glans and thus increases the difficulty in keeping the penis clean if such be desired by the patient

Balanoposthitis Balanoposthitis is an inflammation of the preputial cavity The membrane becomes reddened mottled and moist The epithelium comes off in patches leaving irregular excoriations which ulcerate and discharge pus There is a burning soreness with itching intensified by urination Shallow ulcerations may occur In the severe forms of the disease there are fever and inguinal adenitis In a chronic balanitis of phimosis the mucous surface of the prepuce is granular and may become condylomatous

In localized perimeatal balanitis of the tight meatus the inflammation may extend to involve the frenum In time the meatus becomes so dense that the lesion cannot be differentiated from carcinoma except by biopsy

Balanitis is obvious The diseases to be distinguished are herpes progenitalis lymphogranuloma inguinale chancroid chancre tuberculosis and carcinoma

Paraphimosis This condition is characterized by a tight retracted prepuce which will not return over the swollen glans At times two folds appear the fold nearer the root of the penis containing the constriction which ulcerates

Before the edema becomes fixed the prepuce can be replaced in two stages thus Evacuate the edema from the glans and foreskin to the subcutaneous tissue of the shaft of the penis by squeezing it from the glans toward the shaft Then seize the penis behind the paraphimosis in the fork between the index and middle fingers of the two hands With each thumb make pressure on the glans laterally not from before backward pressing the remaining edema slowly from the glans and gently urging the glans into the ring of tight foreskin It slips back suddenly If the prepuce cannot be reduced the constricting ring must be cut

Gangrene of the penis and scrotum may occur as the result of a virulent infection of the skin and subcutaneous tissues of the debilitated and the aged man

Tuberculosis Tuberculosis of the skin of the penis is said to occur by contact inoculation either orally or genitally Either of these modes of infection must be rare Sexual relations continue between spouses during the early years of renal tuberculosis without resultant inoculation The disease may spread locally into the anterior urethra but no further unless stricture results

Lupus and the other forms of cutaneous tuberculosis may appear on the penis

Anal epithelioma as the disease progresses ulcerates discharges foul irritating putrid material bleeds on the slightest trauma and causes severe pain and tenesmus on defecation At its various stages anal carcinoma may resemble anal fissure lymphogranuloma venereum tuberculous and syphilitic lesions A biopsy should be performed to prove the diagnosis

The prognosis is bad regardless of the treatment instituted Often these lesions develop into large ulcerating sloughing lesions which cause an indescribable degree of severe and constant pain The tumor ulcerates widely, into the pelvic viscera and often into the bladder The malodorous blood discharge and the widely ulcerated perineal region are unavoidable despite any known treatment

THE MALE GENITAL ORGANS

THE PENIS

Congenital Malformations Either the absence or the doubling of the penis is very rare The penis may remain rudimentary *If the lips of the slitlike urogenital opening* on the undersurface of the penis fail to fuse anywhere along their extent hypospadias results this is a common occurrence in false hermaphroditism simulating the female type

Priapism *Priapism may be defined as persistent painful abnormal erection* of the penis without sexual desire It exists in three forms (1) the purely neurofunctional priapism (2) that which results from disturbances in the local circulation and (3) that of unknown causes The first form is observed after injury of the nervous channels as in fractures and dislocations of the vertebral column involving *compression of the spinal cord in tuberculosis or tumors of the spinal cord in* tabes dorsalis and occasionally in other diseases of the central nervous system

Priapism of local origin is observed in cavernitis due to gonorrhea to vesical stone or to phagedenic ulcer of the prepuce in the presence of neoplasm of the penis which has invaded the corpora cavernosa and after trauma with hematoma in the corpora cavernosa There is also priapism in leukemia hemophilia typhoid fever tuberculosis sepsis and acute articular rheumatism Priapism is often accompanied with severe pain It may become complicated by gangrene

Chordee When the urethra is inflamed the exudate may *involve the corpus* spongiosum surrounding it and prevent it from expanding In erection the organ assumes a downward curve a condition designated as chordee It disappears with the subsidence of the inflammation

Diseases of the Skin and Subcutaneous Tissues of the Penis *The skin of the penis is thin loose free from hair except at the root* and has beneath it some fibers of the dartos muscle The skin projects over the glans forming the prepuce and is attached to the neck or collum glandis and underneath as far forward as the urethra forming the frenum The connective tissue beneath the skin is *loose and* free from fat A fibrous sheath (Buck s fascia) surrounds the corpora cavernosa and corpus spongiosum and binds them together

The lymphatics of the prepuce and skin drain into the inguinal nodes those of the glans empty into the nodes just inside the abdomen Since the lymphatic radicles anastomose at the root of the penis a lesion on one side of the organ may involve the lymphatic nodes in the opposite inguinal region

There are sebaceous glands in the skin of the penis near the skin of the scrotum which may *become infected red and swollen and* discharge a tiny drop of pus Cysts may occur in these glands A chancre on the skin over the shaft of the penis is rare Viral lesions caused by the psittacosis lymphogranuloma venereum group rarely occur on the penile skin

Herpes Progenitalis Herpes progenitalis is a vesicular eruption occurring over the genitalia of men and women It is caused by the herpes simplex group of viruses It occurs more commonly on the penile skin than on the genitalia of women *It* often follows sexual intercourse

cavernosa may occur It affects only the laterodorsal terminal two thirds of the erectile bodies The condition is of unknown origin

The disease begins with a mild pain on erection or with a painless deviation of the erect penis The pain may be short lived and without treatment it will cease within a year The deviation on erection upward or to one side increases for a time and then often tends to grow less The patient ceases to be alarmed and accommodates himself to the permanent deflection Those who have extensive lesions may be unable to have erections Flaccidity of the corpora distal to the lesion is frequent and often constitutes the chief complaint

The lesion may be so small as to be impalpable in the flaccid penis and is made apparent only by the deflection on erection When the lesion is palpable it is hard and flat and somewhat irregular and is within the corpus

Fracture of the Penis and Extravasation of Urine Fracture or rupture of the corpora cavernosa may occur from violence The extravasated blood is absorbed and the laceration heals with a scar In erection this part does not expand hence deformity and distortion with interference of function may result

Urine may invade the penis beneath the fibrous sheath but does not invade the glans because the sheath stops at the *collum glandis* (see Perineal Fascia and Muscles p 393)

Benign Tumors of the Skin of the Penis Papillomas of the skin of the penis the so-called venereal warts appear within the tight preputial cavity or in the female within the vulva They may attain an enormous size and become malignant All urethral papillomas should be suspected of malignancy

Condylomata lata broad masses of papillomas about the moist creases of the penis the scrotum and especially the anus are usually due to syphilis

Lipoma may occur in the perineum where it forms a bosselated ovoidal mass

Malignant Tumors of the Penis Cancer Epithelioma or a squamous cell carcinoma of the penis is a slowly progressive tumor which often does not metastasize readily The average age at the time of the first symptom is 50 years No authentic instance of the disease has been reported in a Hebrew circumcised at birth Jewish women rarely have carcinoma of the cervix These two observations may indicate that here are two diseases not common to Jews At least the value of circumcision as a preventive of carcinoma of the penis will have to stand on its own merits it does not receive impetus by the citing of Jewish records It is in non Jewish persons that the disease appears most frequently There is no doubt however that a long or a tight prepuce is present in most of those who have carcinoma of the penis

The common situation of cancer of the penis is about the corona of the glans The lesion may be papillary The tumor becomes infected early ulcerates and sloughs Penile cancer usually metastasizes uniformly to the lymph nodes Occasionally a highly malignant lesion will metastasize to the bones and to the lungs

The tumor often is hidden from the patient s view by the tight prepuce The complaints are referable to the infection In some instances the initial symptom is a lump under the prepuce or the presence of a bloody discharge from the phimosis

On examination not infrequently it is necessary to make a dorsal slit in the prepuce in order to secure sufficient retraction for an adequate inspection of all the parts The appearance of the tumor and its palpable features is variable from a small lump to a large extensive fungating ulcerating lesion The inguinal lymph nodes are often palpable The enlargement of these nodes may be due to the malignant disease or to infection

The venereal wart condylomas granuloma inguinale and tuberculous lesions resemble cancer in some instances so closely that there is no safe criterion for the diagnosis except biopsy Involvement of the lymph nodes can be proved by biopsy If the corpora are definitely invaded with carcinoma it must be assumed that the

or the scrotum Multiple tuberculous nodules have occurred in the corpora cavernosa and corpus spongiosum

Cutaneous tuberculosis situated *on the external genitalia of men has to be dis*tinguished from soft chancre Soft chancre in men may occur on any part of the anorectal skin

Soft Chancre There are three infections of the external genitalia and anorectal regions of both men and women which are characterized by ulcerations and inguinal lymph node involvement and abscess formation (1) soft chancre (2) granuloma inguinale and (3) *lymphopathia venereum The similarity of manifestations of* these diseases and in the terminologies is confusing (see Diseases of the Female Genitalia p 440)

A soft chancre results from an infection by the bacterium *Hemophilus ducreyi* (Ducrey s bacillus) *Granuloma inguinale is caused by a bacterial infection the specific* bacterium is unknown but it characteristically forms Donovania granulomatis Lymphogranuloma venereum is caused by a filtrable virus

The soft chancre or chancroid is caused by *Hemophilus ducreyi* This organism appears in the purulent discharge from the soft chancre or chancroid of a venereal disease for which it is responsible The morphologic structure of the organisms in local lesions is characteristic chains of small gram negative bacilli occur in strands A saline suspension of killed *Hemophilus ducreyi* serves as a good antigen for diagnostic cutaneous tests

Soft chancre with associated abscess in inguinal lymph nodes is perhaps 15 times as common in men as in women It has been suggested that the causal agent *Hemophilus ducreyi* may be carried in the vagina without producing symptoms or ulcerations It is transmitted by sexual intercourse However infections may occur through contaminated towels and instruments uncleanliness and vaginal or urethral discharges *In women chancroids occasionally occur round the vestibule on the* labia and occasionally on anal tags

The incubation period is up to 5 days at the end of which a small reddish macule appears on the genitalia which soon becomes a pustule This ruptures to form a circumscribed ulcer with sharply defined irregular undermined edges and a gray necrotic base The ulcers are multiple as a rule extend peripherally and coalesce to form large irregular ulcers with surrounding inflammatory areolae which are circular or oval rather soft and covered with an abundant purulent secretion These ulcers are extremely painful and tender The inguinal lymph nodes are swollen and tender and are termed *buboes*

The diagnosis of uncomplicated chancroid is based on the identification of *Hemophilus ducreyi* on smear from a characteristic ulcerative lesion a positive reaction to Ducrey vaccine absence of *Treponema pallidum* on darkfield examination and negative Wassermann and Frei reactions Failure to demonstrate Ducrey bacilli on smear does not necessarily rule out the disease In lesions which are contaminated or treated with caustics or of long duration with sloughing areas it is difficult to obtain the organism

Reaction to Intradermal Injection of Ducrey Vaccine The reaction following the intradermal injection of 0 1 ml of Ducrey vaccine is specific At the end of 48 hours a papule 10 mm in diameter surrounded by a zone of erythema 5 mm wide denotes a positive reaction A negative reaction leaves little to be seen at the site of inoculation The reaction becomes *positive between 8 and 15 days after the appearance of the local* lesion A negative reaction after this time rules out a chancroidal infection in about 95 per cent of cases A positive reaction may mean that the patient has a chancroidal lesion at the time the test is performed or that he has had a chancroidal lesion at one time The reaction probably remains *positive throughout the patient s life,*

The disease may yield to treatment with sulfonamides

Fibrosis of the Sheath of the Corpora Cavernosa (Peyronie s Disease) In men between the ages of 30 and 60 years fibrosis of the [illegible]

A slight degree of stricturing causes no symptoms and requires no attention unless the patient has a chronic urethritis such as occurs in gonorrhea In some instances a gonorrheal infection may not be necessary for the origination of the urethritis A simple urethritis with discharge so profuse as to be mistaken for gonorrhea may be due to a strictured meatus not necessarily tightly strictured and disappears on correction of the stricture Some aging men who have symptoms similar to those resulting from a large prostate gland are relieved by meatotomy

Small strictures may interfere enough with the discharge of urine finally to injure the kidneys sufficiently to cause renal insufficiency which may become manifest at any age sometimes in youth sometimes not until old age

Strictures of the posterior urethra of congenital origin are often caused by valve like structures which have definite relationship to the superior or the inferior crista of the verumontanum The symptoms appear in infancy in childhood or in early adult life The stream is dribbling and the bladder distended when retention is great but when the retention is slight the bladder is not distended and the symptoms may be obscure As the result of the retention of urine soon there is an infection of the upper part of the urinary tract and perhaps finally renal insufficiency

The patient often has lost weight and is pale There may be dribbling of urine as if enuresis were present Patients who have had symptoms for a long time may be dwarfed and infantile as is observed in so called renal rickets

There is no interference to the passage of a catheter which usually reveals the presence of residual urine An intravenous urogram discloses the impairment of renal function and the dilatation of the upper urinary organs If renal function is impaired an hour or two must elapse before a satisfactory film can be obtained The valve is identified through a urethroscope It may sometimes be seen by cystoscope

Epispadias is rare The urethra lies ectopic as a groove for varying distances along the top of the corpora cavernosa The penis is wide and short The urethra of the exstrophied bladder is epispadic and the epispadic urethra without exstrophy is usually incontinent

Hypospadias situated in the balanic or penile urethra is but an arrest of the closure of the urethra in these segments There are varying degrees of lack of closure however and thus these malformations range all the way from pseudo hermaphroditism to simple hypospadias

The urethra opens at the frenum or just back of it in balanic hypospadias and on the underside of the shaft of the penis in penile hypospadias In penoscrotal or perineoscrotal hypospadias the urethra opens on the anterior or posterior face of the scrotum but always beyond the sphincters The urethral opening is often deformed and is congenitally strictured

The penis beyond the urethral opening is deformed The glans is wide and split as it were by the failure of the floor of the urethra to unite This broad glans is tilted downward as though pulled over by the shortened urethra Its undersurface is often marked by an indentation or a little pocket at the site of the lacuna magna The congenitally split prepuce divided on the ventral surface is piled up on the dorsum in irregular folds

The tight meatus may cause urinary frequency or infection but the essential symptom of hypospadias is that the patient is ashamed of the deformity Owing to the penile incurvation if the meatus is too far back on the penis to ejaculate into the vagina there is sterility Impregnation may be possible however from a peno scrotal hypospadiac

Nonspecific Urethritis Inflammations of the urethra may be (1) bacterial (2) parasitic (3) traumatic or (4) irritative A distinctive difference between non gonorrheal urethritis and gonorrheal urethritis is that gonorrheal urethritis begins at the meatus whereas all other urethral infections generally originate in the prostate

deep pelvic chain of lymphatic structures is involved and that whatever treatment is considered is likely to be only palliative

Amputation should be done in all cases for if metastasis is not present amputation will cure the condition and if it is present, the procedure will get rid of a badly infected odorous primary tumor

THE MALE URETHRA

(*Urethra Virilis*)

The male urethra varies greatly in length and therefore only average measurements can be given In the adult its length is about 8 inches (20 3 cm) of this 1¼ inches (3 2 cm) is prostatic ⅕ inch (0 5 cm) being in the bladder wall ⅖ inch (1 cm) is mebranous a little more than 6 inches (15 2 cm) is penile Its caliber varies being smaller at the meatus and in the membranous portion than elsewhere The meatus admits a No 24 French sound (often larger) the membranous portion a No 26 to a No 28 The prostatic portion is the largest admitting a No 32 sound The bulbous portion is almost as large admitting a No 30 to a No 32 Therefore a sound which passes the meatus should find no further obstruction The fossa navicularis just beyond the meatus is larger than the rest of the urethra

If urine begins to flow when a catheter is introduced about 8 inches (20 3 cm) it is safe to assume that the urethra is of normal length If the bladder is distended as soon as the catheter passes the membranous portion about 6¾ inches (17 2 cm) from the meatus urine may flow In hypertrophy of the prostate the prostatic urethra is much increased in length and it may require a catheter 10 to 12 inches (25 4 to 30 5 cm) long to draw urine If a sound is stopped by a stricture less than 6 inches (15 2 cm) from the meatus it is anterior to the triangular ligament The stricture is termed an *anterior stricture* Strictures where the subpubic curve is lowest that is in the bulbomembranous region are termed *posterior deep strictures* which are the commonest form Strictures of the prostatic portion of the urethra are almost unknown

The urethra contains small mucous glands (glands of Littre) opening on its surface and small pockets or recesses called the lacunae of Morgagni The glands of Cowper open into the bulbous urethra just in front of the triangular ligament The racemose glands of the prostate open into the sides of the floor of the prostatic urethra and the ejaculatory ducts open near the middle line just in front of the urethral crest (verumontanum)

Urethral Muscles There are two sets of muscles in connection with the urethra one set might be said to aid in expelling the urine and the other in retaining it The expulsors are the longitudinal and circular fibers surrounding the urethra just outside the mucous membrane and the accelerator urinae or *bulbocavernosus* muscle The sphincters are the *compressor urethrae* or *external sphincter* muscle and the internal sphincter at the neck of the bladder

Malformations Important malformations of the male urethra and its accessory glands are of rare occurrence and consequently not all of them will be discussed

Absence of urethra is associated with absence of penis and also may exist when the penis is present There is complete retention of urine and the infant dies unless the bladder drains through urachus or bowel Death usually ensues within a week or 10 days *Atresia* occurs either at the meatus or at the prostatic urethra The condition is usually fatal if some arrangement cannot be made to get rid of the urine

Congenital stricture at the meatus may be only a slight narrowing of the orifice due to a fold of mucosa that elevates the floor of the canal Such a constriction is of no significance unless the patient contracts gonorrhea or until instrumentation becomes necessary The stricture may be so small that only the finest probe will enter The narrowing may be at the meatus or at the inner end of the fossa navicularis or both

Good evidence does not exist of any degree of natural immunity to gonococcal infection The girl child is the most susceptible If any degree of acquired immunity exists it is not measurable There is no infection immunity such as exists in tubercu losis and syphilis That is fresh infections may be superimposed on more chronic ones One attack of the disease does not protect from future reinfection

The incubation period of gonorrhea varies from 2 to 7 days The most accurate information concerning the incubation period of this disease has been obtained from experimental inoculations Under these conditions the urethritis is evident within 2 to 5 days

Acute Gonorrheal Urethritis The initial symptoms of an acute gonorrheal in fection are those of acute anterior urethritis There are a teasing sensation a muco purulent discharge and slight inflammatory swelling of the lips of the meatus In from 1 to 3 days the meatus becomes swollen the discharge thick profuse and purulent and urination and erections are at least uncomfortable and perhaps painful

The inflammation of the urethra with a second attack of gonorrhea may be so mild that it scarcely invades the submucosa The discharge is mild erections are not painful If the infection is unusually intense hematuria may occur as well as urethrorrhagia from bleeding of the anterior urethra

Examination reveals mucopurulent urethral discharge The diagnosis is estab lished by positive smear and culture for the gonococci

The complications of acute urethritis are abscess of the anterior urethral glands periurethral abscess cowperitis spongiitis and cavernitis balanoposthitis and lymphangitis and lymphadenitis These complications can rarely be separately identi fied clinically since small abscesses occur as the result of obstruction of crypts and glands as a part of the acute infection and are important in the production of the symptoms of the disease The most frequent form of periurethral abscess is the frenal abscess It scarcely ever abates without suppuration often requiring incision After incision it may continue indefinitely to discharge a secretion containing gonococci

The less frequently occurring complications may be enumerated (1) Peri urethritis along the pendulous urethra occurs as small hard nodular points of tenderness Introduction of a sound may be necessary in order to palpate them These points too harbor the gonococci (2) Abscess of Cowper s gland is not recognized until the abscess has burrowed along the perineal fascia and its reflection and presents itself at the penoscrotal angle where it bursts or is incised The original focus can be diagnosed by finding that the cavity runs back to the membranous urethra and not to the prostate (3) Inflammation of the erectile tissues other than the brief pains of chordee is rare (4) A lymphadenitis originating from secondary infection from other pyogenic organisms is frequent and in rare instances causes suppuration

The diagnosis is made by stained smears which are microscopically studied for the presence of intracellular gram negative cocci

There is evidence of a gonococcal complement fixation antibody which is acquired slowly after the onset of the infection The complement fixation test in gonococcal disease has not attained general acceptance

Acute Posterior Urethritis and Prostatitis Posterior urethritis and prosta titis are always a part of an acute gonorrheal urethritis As the infection varies in degrees of severity so do the manifestations of its *urethral distribution vary*

The symptoms of posterior urethritis and prostatitis are frequent urgent and painful urination with terminal hematuria There is pus in the second or residual urine The pain of the second infection or the first mild infection is no more than an itching sensation referred to the end of the urethra Posterior urethritis with a first severe attack of gonorrhea is likely to be intense As the muscles contract to expel the last drops of urine the resultant pain is often a series of spasms that may

The types of *bacterial urethritis* are (1) gonorrheal (2) urethritis due to an indwelling catheter which disappears when the catheter is withdrawn though a mild prostatitis is likely to persist (3) urethritis by extension of pyogenic or tuberculous infection originating in the prostate and (4) metastatic urethritis occurring in a canal damaged by previous inflammation Spontaneous severe urethritis may develop when uncontrolled diabetes exists

Parasitic urethritis may occasionally be caused by trichomonas Trichomonas infections are common in the male urethra Feo found 144 infected men in a group of 926 Separating this group into white and Negro men the percentage incidence was 12 and 16 5 respectively Of the 926 men examined 246 were classed as having nonspecific urethritis However in all of these the urethritis was not due to trichomonas The percentage incidence of nonspecific urethritis which may be attributable to *Trichomonas vaginalis* was 36 9 The entire group of *Trichomonas vaginalis* positive men was relatively free from symptoms A discharge may be noted which is characteristically small in amount thin in consistency and of a dirty white color Microscopically this discharge shows a few epithelial cells and a moderate number of pus cells and trichomonads Some of the stained smears were similar to vaginal ones from cases of *Trichomonas vaginalis* vaginitis as to number of trichomonads and types of bacteria The male is the important transmitter of *Trichomonas vaginalis* infection while the female eventually becomes a reservoir of infection

Traumatic urethritis is due to rough passage of instruments urethral stone foreign body or cauterizing injections The more serious and complete stricturing of the urethra is often due to trauma as the result of external violence

Irritative urethritis is due to the excretion of irritating substances in the urine for example, cantharides turpentine arsenic and potassium iodide Irritation from urethral instillations of caustic substances such as silver nitrate solutions may be severe and serious

The symptom of nongonorrheal urethritis is a discharge of pus from the meatus This symptom is mild and inconsequential as compared to the syndrome of acute gonorrhea In diagnosis the absence of gonococci must be proved by urethral culture on the proper media

Posterior Urethritis The lesions of chronic posterior urethritis are (1) diffuse inflammations (2) granulomas (3) cysts (4) deformities due to operation or to rupture and scarring of a prostatic abscess and (5) verumontanitis and utriculitis

The symptoms of chronic posterior urethritis are often mild and manifested by a lack of erection premature ejaculation painful ejaculation and pain in the perineum in a testicle or in the sacrum These symptoms are like those often attributed to verumontanitis for instance absence of ejaculation painful ejaculation discomfort of various types in the perineum and various reflex pains

The diagnosis is established by endoscopic examination

Urethral Gonorrhea Urethral gonorrhea is caused by the gonococcus which belongs to the family Neisseriaceae genus *Neisseria Neisseria gonorrhoeae* is a gram negative coccus which is the type species To the same family and genus belong also the meningococcus and several nonpathogenic inhabitants of mucous membrane

Neisseria gonorrhoeae is the causative agent of gonorrhea and of ophthalmia neonatorum The organisms appear in the exudate of acute gonorrhea as diplococci flattened or slightly concave and crowded together in pairs

Gonorrheal infection occurs spontaneously only in man The gonococcus causes infection of the genitalia and the genital tract generally and may give rise to conjunctivitis cystitis proctitis hepatitis and stomatitis The septicemic manifestations of gonococcal infection are endocarditis arthritis and meningitis The chemical constitution of the gonococcus indicates that the organism contains nucleoproteins and polysaccharides similar to those of the meningococcus

may progress to complete pathologic amputation of the penis This too is rare and the diagnosis is not made

Stricture As chronic urethritis heals a scar is formed A stricture is the scar that results Strictures external to the bulbous urethra are termed *anterior* strictures Those beyond this point are *posterior* or deep strictures Strictures are classified as *inflammatory* and *traumatic*

Inflammatory Strictures Severe strictures result from gonorrhea or are complications thereof Usually only mild strictures result from nongonorrheal inflammation

Strictures as the result of gonorrhea are more frequent after virulent infections than after mild infections For instance the more intense the chordee the more frequent and violent the relapses and the longer the gonococcus can be found in the discharge the greater is the probability of strictures

Inflammatory stricture may result from scarring about the obstructed and chronically inflamed glands or lacunae The overdilatation of a stricture may be followed by a more severe stricture Suppuration may follow overdilatation or may occur spontaneously The direction that a suppurative process takes is determined by the fascia If a prostatic abscess bursts backward or suppuration following overdistention occurs in the same place and does not drain forward into the urethra the pus either reaches the ischiorectal fossa and appears at the surface in front of or alongside of the anus or is confined by the fascia

The following enumeration lists other suppurative processes which complicate stricture (1) The suppuration of prostatic abscess especially if due to tuberculosis often reaches the surface in the perineum or may extend forward to the scrotum and up into the groin between the penis and the spermatic cord Thence it spreads upward and outward toward the loin beneath the deep layer of the superficial fascia (2) Suppuration in Cowper s gland extends forward and passes along the urethra to the penoscrotal angle and surface of the scrotum (3) Suppuration from the bulbous urethra usually breaks directly into the perineum whence it extends to the ischiorectal fossa scrotum groin pubes or anterior abdominal wall (4) Suppuration from the penile urethra bursts directly through the skin of the penis (5) Inflammation and dilatation of the bladder the ureters and the kidneys are late results of strictures A stricture unaccompanied by sclerosis or spasm of the vesical neck does not cause partial retention of urine Stone in the bladder is rarely the result of stricture

Traumatic Strictures The penile portion of the urethra may be cut torn or broken by external violence A crushing force applied to the perineum causes a stricture at the bulbomembranous junction by bringing the urethra sharply into contact with the undersurface of the pubis crushing it beneath the sharp edge of this structure Falling astride a fence or a fence post may cause a traumatic stricture in the posterior portion of the urethra These injuries to the urethra may be overlooked by the patient if they do not give rise to immediate hemorrhage or urinary retention The prostatic urethra becomes strictured when torn by disruption of the pelvis Trauma originating from within such as ulceration from stone foreign body or retained catheter or injury by the sharp point of an instrument usually affects the bulbomembranous junction of the urethra Stricture at the neck of the bladder results from chronic prostatitis and transurethral resection of the prostate

Within one year after a severe inflammatory process or trauma most patients will have strictures with manifest symptoms A few will show no symptoms until after 5 years In an occasional patient the onset may be deferred for 15 years or even longer Organic stricture may exist for years producing no symptoms and entirely unsuspected by the patient

The initial symptom is usually the presence of pus and heavy thick shreds of pus in the urine After stricture formation which immediately follows gonorrhea the urethral discharge does not cease

last for a few minutes after urination or may continue in the tonic spasm of strangury during the short period between successive urinations In these patients urination is genuinely agonizing

Toward the end of the acute period of gonorrhea (2 months) the patient even if untreated with antibiotics or chemotherapy has no symptom other than a drop of pus at the meatus when he awakes in the morning (the morning drop) This drop of pus may be so mucoid that it may stick the lips of the meatus together

The diagnosis is established by positive smears and cultures

The complications of acute posterior gonococcal urethritis are prostatic abscess *vesiculitis epididymitis cystitis* and occasionally pyelonephritis and peritonitis

The distinction between acute prostatitis in which miliary abscesses have not yet appeared and *prostatic abscess* cannot be made clinically The gonococcic prostatic abscess and *vesiculitis* have no distinguishing features from any other etiologic types of these diseases When there is *epididymitis* there is vesiculitis The *cystitis* of gonorrhea is a trigonitis It is an extension of the mucosal inflammation of the posterior urethra A general cystitis may occur in gonorrhea if there is a retention of urine

A nongonococcal pyelonephritis may occur in the course of gonorrhea

Chronic Gonococcal Anterior Urethritis When the disease lasts for more than 2 months the gonococci diminish in number in the pockets in the urethral wall to be replaced by other bacteria which keep up a mild inflammation If stricturing and poor drainage prevail in a debilitated patient anaerobic bacteria may cause periurethral gangrene

The most persistent form of chronic anterior gonococcal urethritis is caused by a frenal abscess The urethra may become free from gonococci only to be reinfected from this source The course of a chronic gonococcal urethritis is often a series of exacerbations of the acute process Apparent improvement or cure is followed by exacerbation seemingly caused by indiscretion by reinfection of the surface of the urethra from an infected focus

The diagnosis is established by positive smears and cultures

Chronic Gonococcal Posterior Urethritis The clinician can rarely obtain a positive smear for gonococci from the prostate or the vesicles The bacteriologists however can often get positive cultures

The inflammation loses its characteristic irritability and the urethritis tends to run a milder and more even course after the gonococci have disappeared Many of these patients have a posterior urethritis by secondary invaders Although some of these patients have not had gonorrhea it is their lot to have an irritable prostate which has been treated for gonorrhea which they did not have

Prognosis in Urethral Gonorrhea The foregoing description of gonorrhea in the male is becoming out of date since the advent of chemotherapy and antibiotics (duracillin) The following prognosis applies to those who have not been thus treated

The average man should recover from gonorrhea in 3 months if he has no complications such as stricture prostatitis epididymitis or vesiculitis If the infection is complicated by a frenal abscess the course may be indefinite It is most unusual for the gonococcus to persist in the male Prior to use of antibiotics many patients recovered from gonorrhea without or despite treatment

Chronic gonorrhea in women may almost last indefinitely

Amicrobic Urethritis In France there has been described an increased incidence of amicrobic urethritis in men that has proved resistant to penicillin and sulfonamide compounds Harkness has insisted that this infection is caused by a virus This form of urethritis may be associated with prostatitis and epididymitis

Syphilis Chancre of the urethra as a rule is not recognized Chancre has not been described in the deep urethra Gumma occurs in the corpora ca a and

tinence Diverticula of the penile urethra may produce a swelling of the ventral portion of the penis when the patient voids

Visualization of the urethra by cysto-urethrography is the best method of diagnosis Radiopaque substances injected into the urethra clearly outline the diverticular sac

Tumors The neoplasms of the male urethra arise from the epithelium Tumors arising from other elements such as sarcomas are rare

Benign Tumors Benign papillary tumors occur in any portion of the urethra but are commonest in the posterior portion These tumors of the male urethra usually are small delicate neoplasms Histologically they are composed of epithelial cells mitotic figures are not present and there is no evidence of invasion or other signs of malignancy Initial hematuria is the most frequent evidence of urethral papilloma Tumors situated in the posterior part of the urethra may cause sterility by occlusion of the ejaculatory ducts The diagnosis is established by endoscopic examination and biopsy

Malignant Tumors Carcinoma is the commonest primary malignant tumor of the male urethra This lesion occurs most often in men between 40 and 50 years of age

Cancer may arise in any portion of the male urethra There has been no significant difference in its frequency in the anterior and the posterior portions of the urethra Histologically these tumors usually are epidermoid carcinoma or adeno carcinoma Primary carcinoma of the urethra metastasizes late in the disease

Slow stream frequency dysuria and hematuria are symptoms of primary carcinoma of the urethra Considerable infection seems to be a feature of these neoplasms and abscess formation and fistulas are common

Diagnosis is established by biopsy Early diagnosis of urethral carcinoma is difficult Symptoms are so suggestive of stricture of the urethra that many patients with urethral cancer have been treated for stricture Tumors of appreciable size can be felt along the urethra Any progressive narrowing of the urinary stream associated with a palpable mass along the course of the urethra should suggest carcinoma

THE SCROTUM

Anomalies Anomalies of the scrotum include underdevelopment of the scrotal sac associated with undescended testicles and infantilism Bifid scrotum may be associated with exstrophy epispadias and hypospadias

Infections Many of the conditions affecting the skin also affect the scrotum However the scrotum is peculiarly resistant to infections

Gangrene The so called idiopathic gangrene of the scrotum is of sudden onset without apparent cause Symptoms begin suddenly in an apparently healthy man The scrotal skin becomes tense red and shiny The patient becomes severely toxemic in appearance Gangrene appears and spreads rapidly and soon the skin sloughs away The process extends beneath Colles fascia and may spread into the groins and onto the abdominal wall The testicles are left exposed Even after extensive involvement the process ceases suddenly as it commenced granulation appears and the scrotal skin regenerates quickly

Gangrene is differentiated from phlegmon by the absence of history of disease causing extravasation of urine Phlegmon does not slough extensively One of each 5 who have the disease dies

Elephantiasis Elephantiasis of the scrotum is endemic in many southern regions The disease is due to infestation by a nematode worm *Wuchereria bancrofti*

The symptoms of filariasis are episodes of lymphangitis lymphadenitis and swelling of the scrotum Repeated attacks of the disease may result in enormous scrotal enlargements Except for this deformity the physical condition of the patient is unaffected

Frequent micturition often painful is due to straining to overcome the urethral resistance, prostatitis and cystitis If there is pyelonephritis, the frequent urination may be a true polyuria

If a stricture becomes congested it closes and acute retention of urine ensues If this retention is unrelieved the progression is like that of an acute prostatic retention from benign hypertrophy

As the stricture contracts the urethritis grows worse and sooner or later produces a morning discharge (gleet) The morning discharge gets better or worse according to the degree of acidity of the urine and the amount of sexual indulgence As the stricture tightens the stream of urine is small and irregular Erection is sometimes imperfect and painful

The pain on urination in stricture is due to prostatitis cystitis or retention It is *perceived deep in the urethra in the perineum at the point of stricture or near the* glans penis

When gross hematuria occurs it is often the only symptom of stricture noticed by the patient The bleeding comes from a shallow ulcer and may be profuse The blood usually continues to drip after the close of micturition (urethrorrhagia)

DIAGNOSIS The diagnosis of strictures is made only by those trained to use the bougie and the sound (1) In stricture of the pendulous or scrotal urethra the bulbous bougie either fails to pass the strictured point or passes with a jump and is caught again by the stricture on withdrawal (2) Stricture at the bulb is diagnosed by use of a sound A sound distinguishes stricture from spasm by the fact that when it has entered the stricture it is grasped on withdrawal Blood follows withdrawal of the sound (3) Spasmodic stricture is a symptom not a disease It is a subjective spasm created by an unduly irritable muscle because of urethritis or prostatitis Urethral spasm is often a postoperative complication (4) Traumatic strictures contract rapidly and are usually incurable The more extensive a stricture the more irregular its surface and the denser the cicatricial tissue composing it the more difficult will be its treatment and the more dubious its cure

If death results from a stricture it is from septicemia obstruction of urine and renal failure or periurethral gangrene When the patient is debilitated sudden death may occur after the passage of a sound

Urethral Stones Vesical calculi may pass into the urethra and become impacted along its course Prostatic calculi occasionally erode through the mucosa and obstruct the urethra Primary urethral calculi may form behind a stricture or within a diverticulum Urethral calculi cause varying degrees of obstruction depending on their size

Patients who have urethral calculi often are unaware of the presence of the stones The diagnosis frequently is made by the passage of some instrument into the urethra Characteristic grating then may be felt This sensation may be obtained at times when tough old strictures of the urethra exist so that stone is suspected although not present Stones within urethral diverticula are detected by roentgenologic examination

Diverticula Diverticula of the male urethra are uncommon They are classified as congenital and acquired

Congenital Diverticula Diverticula of this type are said to occur most often on the floor of the penile portion of the urethra Their origin is thought to be due to incomplete fusion of that portion of the urethra which is derived from the phallic part of the genital tubercle with the portion of the urethra which is derived from the urogenital sinus

Acquired Diverticula The acquired types of urethral diverticula are due to the presence of stricture or stone abscess formation or trauma The symptoms if present are those of chronic inflammation dysuria dribbling and incon

The testis is covered by peritoneum which is prolonged at its upper and lower ends The lower end reaches down to the internal ring and later contains fibrous and muscular tissue and passes through the inguinal canal to the lower part of the scrotum it is called the gubernaculum testis It reaches its highest development in the sixth month and its remains attach the testicle to the lower part of the scrotum as the ligament of the scrotum As the testicle descends it is preceded in its descent through the inguinal canal by a fold of peritoneum the vaginal process which forms the tunica vaginalis over the testicle

The vaginal process may not entirely close so that the peritoneal fluid passes down to the tunica vaginalis covering the testicle this is called a *congenital hydro cele* If the opening is large enough for intestine to enter it forms a *congenital hernia* If the opening is closed above usually at the external ring and fluid ac cumulates in the tunica vaginalis it forms an *infantile hydrocele* If a portion of the vaginal process persists somewhere along the spermatic cord between the internal ring and top of the testis there is an *encysted hydrocele* A hydrocele in the inguinal canal of the female is a hydrocele of the canal of Nuck

Anomalies of the Testicles *Anorchism* or *monorchism* is extremely rare unless there is in addition an absence of one or more of the other elements of the seminal tract The failure to find an undescended testicle on surgical exploration is not prima facie evidence of its congenital absence Likewise *synorchism* is not a good explanation for not finding the second testicle

Polyorchism is rare but when present there is but one vas and the duplicated testicles are joined by the epididymis

The absence of one testicle is rare but when one testicle is lacking there is absence of the homolateral kidney also for there is absence of the whole urogenital fold

In *cryptorchism* the ectopic testicle may be halted at any point of its normal descent It may be situated anywhere throughout the length of the inguinal canal In infancy retention in the canal is common Retention frequently occurs at the external inguinal ring in the groin and in the proximal portion of the scrotum

The ectopic testicle becomes sterile because of the slightly higher temperature of its bed in the groin or inguinal canal than of the normal situation in the thin walled scrotum Puberty is the accepted dividing line in determination of normal development of the testicle It is expected that placement of the testicle in the scrotum before puberty will result in normal testicular development in size and function Retention of the testicle above the external inguinal ring after puberty is accepted as proof that the testicle has failed to attain normal size and spermatogenic function

When both testicles are ectopic there is no spermatogenic function although the hormones that determine the development of the secondary sexual character-istics and preside over the growth of the prostate and seminal vesicles are produced in undiminished quantities The secondary characteristics of sex develop normally

The undescended testicle is much more liable to malignant growth than the normally situated testicle In almost every instance if the testicle does not descend there is a congenital *hernial sac* above the ectopic testicle

Infections of the Testicles Direct extension of inflammation from the epididy mis accounts for most instances of local inflammatory disease of the testicles Blood borne infections of the testis occur in pyogenic generalized infections mumps syphilis the rickettsial fevers and typhoid fever

Epididymo orchitis occurs as a complication of injuries and operations on the bladder and prostate In severe epididymo orchitis the substance of the testis is destroyed by acute inflammation and abscess formation The swollen scrotal con tents cause considerable pain and discomfort The cord is enlarged and tender along its course giving pain and tenderness in the lower part of the abdomen suggestive at times of acute appendicitis As the scrotal mass increases in size it first is hard and then becomes fluctuant Acute severe epididymo orchitis is obvious

Lymphedema Progressive lymphedema of the extremities and scrotum occurs among men who never have resided in the tropics This condition is the result of obstruction of the lymph channels and like Milroy s disease of young women may be congenital in origin Rarely is there a history of any significant infection or injury

Tumors New growths of the scrotum are not common

Benign Tumors The benign tumors may be enumerated (1) Vascular neoplasms and an occasional accessory spleen in the scrotum are recorded as examples of tumors of congenital origin (2) Sebaceous cysts are common They appear as yellowish round freely movable tumors which vary in size and number The cysts are filled with thick foul debris Usually scrotal cysts require no treatment (3) Lipomas are composed of large lobules of well encapsulated fatty tissue Symptoms result only from weight and pressure (4) Connective tissue tumors of the scrotum are slowly growing small hard neoplasms (5) Myxomas of the scrotum occur as smooth lobulated masses and may attain considerable size These neoplasms are said to transilluminate (6) Tumors of the lymphatic vessels occur as diffuse lesions The skin is edematous indurated and often ulcerated These neoplasms are occasionally malignant (7) *Hemangiomas originating in the subcutaneous tissues* may slowly enlarge They are composed of masses of dilated blood vessels filled with blood

Malignant Tumors Most malignant tumors of the scrotum are epitheliomas

CARCINOMA OR EPITHELIOMA Considerable interest is attached to carcinoma of the scrotum because of its rapid growth high mortality and occupational incidence This was the first recognized occupational cancer

Carcinoma of the scrotum may develop in workers in certain industries as a result of their occupation Chiefly these are chimney sweeps employees in textile industries and men exposed to tar pitch and lubricating oils *Mule spinner s disease* is cancer of the scrotum occurring in men who work in the cotton industry where they are exposed to the carcinogenic agent present in mineral oil used for lubrication This disease has nothing to do with the faithful farm animal the southern mule

Carcinoma or epithelioma of the scrotum is epidermoid carcinoma The lesion starts as a small wart which soon ulcerates The ulcer is hard and indurated and has everted edges As the lesion advances the lymph nodes of the groins become involved and are felt as hard tender masses which also eventually ulcerate

Scrotal cancer metastasizes to the inguinal and femoral lymph nodes Crossed metastasis occurs frequently

The diagnosis is established by biopsy Scrotal carcinomas are highly malignant tumors Even with early excision the prognosis is not good

SARCOMA Sarcoma of the scrotum is rare

THE TESTICLES AND ASSOCIATED STRUCTURES

The Testicles The normal testicles are 1½ inches (3 8 cm) long 1 inch (2 5 cm) wide and ⅘ inch (2 cm) thick If larger they are either hypertrophied or diseased They are firm to the touch If hypertrophied their consistency is not materially altered if diseased they are usually harder than normal If smaller they are usually atrophied and besides the lessening of size are also softer and flabby in consistency

The testicles rest attached at the inner posterior portion of the scrotum and the long axis points upward slightly forward and outward In the presence of hernia and hydrocele the testicle is to be felt for first at the inner posterior aspect of the swelling If not found here the testicle may be palpated anteriorly instead of posteriorly The position of the testicle in the presence of hernia or hydrocele can also be determined by seeing its outline by means of a light placed on the opposite side of the scrotum

Descent of the Testicles *The testicles begin to develop early in fetal life* at about the third month below and in front of the kidneys opposite the second lumbar vertebra

pathic hydrocele is a slow accumulation of fluid within the tunica vaginalis for no known reason The soft tumor of the testicle transilluminates and on needle puncture yields clear straw-colored fluid

Hydrocele must be differentiated from hernia Hernia does not transilluminate gives an impulse on coughing and often can be reduced Spermatoceles contain milky fluid containing spermatozoa and usually are situated at the upper pole of the epididymis Chronic hydroceles with thickened sacs if large and uncomfortable may require excision

Neuralgia True neuralgia of the testicle or cord is an ache constant and mild which often is of long duration The ache or pain may be situated throughout the length of the cord or in the testicle The pain may be centered in the epididymis In instances of severe pain of neuralgic nature in the testicle removal of the testicles will not relieve the pain The operation will make the neuralgia worse

Prolonged sexual desire may produce neuralgia (*stoneache*)

Tumors of the Testicles Tumors of the testicles are both benign and malignant

BENIGN TUMORS Hyperplasia of the interstitial cells of the testicles may occur in children This condition may be accompanied by precocious bodily and sexual development Gynecomastia sometimes is present Adenomas fibromas and hemangiomas have been reported but they are rare Simple cysts of the testis are rare So called benign teratomas have been described but these tumors sometimes are malignant since they have been known to metastasize

Benign tumors of the testis can be differentiated from malignant neoplasms only by microscopic examination

MALIGNANT TUMORS Malignant tumors of the testis are often a most dangerous form of malignant disease

Malignant testicular neoplasms have been reported from birth to old age but they are commonest between 20 and 45 years of age with an average age of about 35 years The association of malignant tumors with cryptorchidism is well established

Friedman and Moore found on examination of 922 testicular neoplasms that 96 per cent of these new growths fell into one of the following categories (1) seminoma (germinoma) (2) (a) embryonal carcinoma and (b) chorio epithelioma (3) teratoma and (4) teratocarcinoma

The *seminoma* is one of the commonest of malignant testicular tumors Seminomas have less malignant characteristics than embryonal carcinomas in that they tend less to invade and destroy adjacent tissues They occur at a later age than other testicular neoplasms *Embryonal carcinomas* fortunately are less common than seminomas *Chorio epitheliomas* of the testis are rare These are small soft hemorrhagic tumors They grow very rapidly and have a poor prognosis Characteristic of chorio epitheliomas are their rapid growth early wide dissemination lack of radiosensitivity and high titer of chorionic gonadotrophin in the urine Most of these tumors end fatally within a short time

Teratomas are bizarre neoplasms which contain differentiated adult structures such as bone cartilage or well formed organs resembling teeth and hair Any organ of the body may be represented in these tumors They are slowly growing neoplasms which constitute fewer than 1 of each 10 new growths of the testicles *Teratocarcinoma* or embryonal adenocarcinoma usually consists of a mixture of teratoid elements in the rete testis but the epithelial elements grow rapidly and soon overgrow the tissue of origin It is a highly malignant tumor

ENDOCRINE TUMORS OF THE TESTES In this discussion it is convenient to consider that a testicular tumor may arise from any one of the four different cellular structures (1) the supporting connective tissue (2) the interstitial cells of Leydig (3) the supporting cells of Sertoli and (4) the germ cells Tumors arising from cellular structures other than the germ cells are scarce Three in every 100 testicular

ORCHITIS IN MUMPS Orchitis in mumps rarely occurs before puberty After puberty it occurs in about 1 of every 5 who have the mumps Severe orchitis is fol lowed by atrophy of the testis *in about one half of the cases*

Orchitis in mumps appears within 3 or 4 days to 1 or 2 weeks after the onset of the parotid swelling The involved testicle or testicles are swollen firm and very tender Epididymitis of mumps may occur alone without orchitis or parotitis and in some instances with very little parotid enlargement

Orchitis in mumps after puberty is a serious condition for testicular atrophy and sterility may ensue if both testicles are affected

SYPHILIS *Testicular involvement* is an early and a constant consequence of syphilis Two forms of syphilis of the testicle are described either a sclerotic or a sclerogummatous *lesion may be present*

Bilateral sclerotic orchitis is characteristic of late syphilis The only symptom present is diminution or loss of testicular sensation

Sclerogummatous syphilitic orchitis tends to localize in the rete and to extend thence to the epididymis and adjacent tunica vaginalis The gumma may be palpated as an irregular insensitive mass on top of the testicle with a hard ridge along one side The opposite testicle is insensitive to pressure

The forementioned findings of a testicular mass with the opposite testicle in sensitive and the history of syphilis in the presence of positive serologic reactions are diagnostic

Torsion The cause of torsion is often unknown to the patient If torsion is more than one half of a revolution and if it is not promptly untwisted gangrene of the testicle and epididymis ensue

Torsion may occur in infants but in most instances perhaps 3 out of 4 it occurs between the ages of 15 and 25 years in the remainder it occurs in adult life It is usually situated on the right side

Slight torsion may recur a number of times before the circulation is sufficiently injured to cause atrophy of the testicle The onset of the pain may be mild or severe Often attacks of mild pain and swelling of the testicle occur and subside to be fol lowed by more attacks thus simulating epididymitis of gonorrhea or recurring trauma

In severe torsion the gangrenous testicle within a few days reaches an enormous size and its sensibility diminishes but pain and fever persist

The diagnosis of complete torsion *of the cord is established by the history* and examination Mild torsion may not be recognized before it becomes complete If mild torsion is diagnosed the scrotum should be incised and the testicle sutured to the scrotum Had the gubernaculum of the testes anchored the testicle tightly to the bottom of the scrotum torsion would not have occurred

Injuries of the Testicles The testicles are protected against injury by their mobility and position Injuries occur from direct blows and lacerations Causes of injury may be enumerated as follows (1) Exposure to roentgen rays may seriously affect the spermatogenic elements of the testis (2) Contusions of the testis result from direct blows These injuries cause severe pain because of edema and regions of hemorrhage within the organ Contusions of the organ may result in hydrocele hematocele and sterility Piercing wounds of the testicle are often serious and are followed by hemorrhage (3) Lacerations of the testis may occur from penetrating wounds of the scrotum Lacerations require suture of the tunica albuginea to pre vent *herniation of the testicular tubules*

Severe injuries of the testicle are accompanied by shock Pain is severe The scrotum becomes enlarged and discolored

Hydrocele Accumulation of fluid within the tunica vaginalis occurs as an acute or a chronic condition Acute hydrocele results from trauma inflammation such as epididymitis or orchitis and sometimes *from*

has been the subject of considerable comment Confusion regarding the value of these tests arises when it is not recognized that the hormone present in the urine in those who have testicular tumors is different from the hormone present in castrates and in women past the menopause The presence of chorionic gonadotropic hormone in the urine of a patient who has a possible malignant tumor of the testicle is an evidence of the presence of a tumor containing chorionic tissue A decrease in the chorionic gonadotropic hormone means removal or regression of chorionic tumor tissue An increase means a spread of the disease or an increase of follicle stimulating hormone with a synergistic effect on the chorionic gonadotropic hormone

Since the reactions to the Aschheim Zondek and Friedman tests often are negative in seminoma and other tumors of high malignancy the value of these tests in diagnosis is limited However the presence of chorionic gonadotropic hormone in the urine means that there is an active tumor somewhere and suggests a bad prognosis The amount of gonadotrophin present in the urine is significant Patients whose urine contains 2 500 or more mouse units per liter are short lived All of those who have 10 000 mouse units in the urine have been proved to have chorioepithelioma

Most teratomas (adult teratomas) without malignant elements are cured by orchiectomy alone Seminomas respond well to orchiectomy and external radiation and many of these tumors probably are cured Embryonal carcinomas and teratocarcinomas have a higher mortality rate and in these neoplasms radical surgical intervention may produce better results Chorio epithelioma generally gives a poor prognosis although an occasional patient with this neoplasm has survived for a number of years

The ten year survival rate for treated patients who had seminoma as reported by Cabot and Berkson was nearly 50 per cent as compared with a ten year survival rate of about 25 per cent for patients with other forms of carcinoma

Tumors of the Testicular Tunics and Spermatic Cord The majority of the tumors of the testicular tunics are of mesoblastic origin and are lipomas myxomas fibromas or mixtures of cell elements Sarcomas occasionally occur All of these are rare tumors

Most tumors of the *tunica vaginalis* are benign A few epithelial tumors have been reported all benign adenomas Other benign neoplasms are lipomas fibromas and myomas All the malignant tumors have been sarcomas The *tunica albuginea* gives rise to fibromas or sarcomas

Tumors arising from the testicular tunics usually resemble testicular neoplasms and are treated as such A few benign growths have been removed by local excision

Tumors originating from the *spermatic cord* are all mesoblastic and usually benign The commonest tumor of the cord is the lipoma

The Spermatic Cord The left spermatic cord is longer than the right hence the left testicle hangs lower The cord is composed of the vas deferens with its artery and veins the spermatic artery with its veins the cremasteric artery and the layers derived from the abdominal wall which extend to become the tunics of the testicle The vas deferens is a small round hard cord lying posteriorly which is palpable through the scrotum

Varicocele Varicocele is varicosity of the veins of the pampiniform plexus of the spermatic cord *Spontaneous varicocele* a mild form of varicocele is a common affection It occurs usually on the left side The deformity makes itself manifest before the age of 25 years A *symptomatic varicocele* may occur as the result of pressure on the spermatic vein on either side or on both sides

Some patients with varicocele have hypochondriac complaints Old men do not complain of varicocele Perhaps there is a slight dragging sensation in the groin On examination the vessels are dilated and feel like a soft tangled cord or a bundle of worms

tumors studied by Friedman and Moore were germ cell tumors Of the germinal cell tumors those arising from the Leydig and Sertoli cells are rare but their clinical manifestations are complex for the Leydig extratubular cells produce androgen and the Sertoli intratubular cells produce estrogen Therefore a condition is originated in which a masculinizing and a feminizing hormone are being produced side by side in the same gland If the Leydig cell secretion androgen is excessive during youth a giant is the consequence If the Sertoli cell secretion is increased the youth is feminine in character

The chorionic cell tumors produce a luteinizing type of gonadotrophin If gonadotrophins are produced in excess estrogen also is produced in excess and gynecomastia ensues It is said that any male who has enlarged breasts suffers from an endocrine imbalance This may be true but often gynecomastia is present in youth without evidence of endocrine imbalance (see Diseases of the Breast Chapter 8) Gynecomastia may be associated with disturbed testicular function observed after trauma cryptorchidism senility severe hepatic damage and the administration of stilbestrol

The excess estrogen in those who have chorio epithelioma of the testes is formed in the tumor itself and can be extracted from it The extract (gonadotrophin) from chorio epithelioma of the testis gives a biologic test similar to that of the pregnancy urine test It is the amount of gonadotrophin present in the urine which is signifi cant Large amounts (10 000 to 100 000 or more units) may be present in the urine from one who has a chorio epithelioma The amount of gonadotrophins present is indicative of the mass of functional tumor cells present

Malignant tumors of the testis metastasize by lymphatic structures along the spermatic cord to para aortic lymph nodes then through the prevertebral chain of lymph nodes through the mediastinum and along the thoracic duct to the left supra clavicular fossa and perhaps to the lung Metastasis to the left supraclavicular lymph node is present in an occasional case Crossed metastasis at a high level may occur Inguinal lymph node involvement occurs when the tumor has involved the scrotal tissue The size of the primary tumor has little to do with the onset of metastasis

Despite the ready accessibility of the organ symptoms of tumor of the testis frequently are not noticed for a long time Large masses may cause dull dragging discomfort The presence of an abdominal mass or backache from involvement of retroperitoneal lymph nodes may be the first symptom of testicular tumor Some tumors have an onset resembling acute epididymitis In many instances trauma brings the tumor to the patient s attention Often the first manifestation of these new growths is a metastatic lesion found on routine physical examination or the presence of a metastatic nodule in a lung revealed by roentgenographic examination or an enlarged left supraclavicular lymph node Hydrocele may be found in association with testicular neoplasm Gynecomastia may be present

The diagnosis is made by biopsy after removal of the testicle for a tumor Biopsy of the suspected testicle is never performed since this procedure is likely to spread tumor cells and lead to local recurrence If it is strongly suspected that a malignant testicular neoplasm is present the entire organ with its tunics intact is removed Difficulties in the diagnosis of these tumors are many Testicular neoplasms have been mistaken for hydrocele hematocele epididymitis orchitis and tuberculosis It is not possible to differentiate benign and malignant tumors by physical examination

Any painless swelling of the testicle is suspected of being a neoplasm The shape of the normal testicle is likely to be preserved with most malignant neoplasms and the mass feels heavy If hydrocele is present the fluid may be aspirated to provide better examination of the testicle The cord usually feels normal

The value of hormonal tests in the diagnosis of malignant testicular neoplasms

is made Orchiectomy should not be performed until treatment with antibiotics has been given a trial

Tumors of the Epididymis Primary tumors of the epididymis are rare They are benign or malignant

BENIGN TUMORS *Spermatoceles* are common They may reach several centimeters in size They contain a milky colored fluid and transilluminate Almost half of the primary neoplasms of the epididymis lipomas adenomas leiomyomas and angiomas are said to be benign Mesotheliomas are nonmalignant neoplasms primary in the epididymis

Benign tumors of the epididymis are small and cause no symptoms The presence of these benign tumors is detected in the course of a physical examination Only by histologic examination is the diagnosis established

MALIGNANT TUMORS These tumors are about equally divided in their origin from epithelium and mesothelium Pain and a solid mass are the manifestations of a primary malignant tumor of the epididymis The diagnosis is established by biopsy

The Vas Deferens The vas deferens descends to the lower end of the testicle and becoming much convoluted forms the globus minor or tail thence it ascends forming the body and finally at the top receiving the efferent ducts forms the globus major or head Between the body of the epididymis and testis is a pocket or depression called the digital fossa Attached to the upper end of the testis is a small flat body which is in front of the globus major Attached to the globus major itself is a small cystic pedunculated growth Both of these masses are known as the hydatids of Morgagni the former is the remains of the duct of Muller the latter is derived from the wolffian body The vas deferens then ascends along the spermatic cord to the internal abdominal ring of the inguinal canal

When the vas deferens leaves the internal abdominal ring it winds around the outer side of the deep epigastric artery and dips down over the brim of the pelvis 1½ to 2 inches (3 8 to 5 1 cm) posterior to the pubic spine It then runs downward and backward on the side of the pelvis In its pelvic course the vas deferens is not often the subject of clinical significance except in cases of undescended testis

Deferentitis Deferentitis is recognized clinically in both acute and chronic forms which often follow gonorrheal infections Also both acute and chronic forms of tuberculous infections are observed

The *acute infection* may be recognized as a thickening of the seminal vesicle which is palpable at the external ring Commonly deferentitis extends from a tuberculous epididymis backward up the vas deferens Muscular trauma at the inguinal canal occasionally excites suppuration

Chronic tuberculous deferentitis is an irregularly distributed lesion palpably knobby and usually most marked in the scrotal portion As in epididymitis it is tender or insensitive in proportion to the acuteness of the infection

The Seminal Vesicles The seminal vesicles are about 2 inches (5 1 cm) long and lie on the bladder above the prostate When normal they are not readily recognized by touch but in disease they are easily felt

Anomalies Anomalies of the vesicles are usually part of some general genital malformation When one of the vesicles is absent the corresponding testicle may yet be present Extreme dilatation of the vesicles is probably always acquired

The ejaculatory ducts may empty into the ureters instead of on the edge of the prostatic utricle In a few cases they have been found to continue forward alongside of the urethra the whole length of that canal to the meatus

Wounds Operative wounds of the ejaculatory ducts are frequent The patency of the ducts is imperiled by prostatectomy

Cysts A gradual dilatation results from inflammation of the vesicles until they become two or three times their normal size and even overlap in the median line Such dilatations have no clinical significance In rare instances one seminal vesicle may become obstructed at its outlet and then it will enlarge and form a tumor about

Tumors Tumors of the cord are rare A lipoma is about the only tumor of the cord It is secondary to hernial lipoma is usually small and is present in an inguinal hernia

The Epididymis The epididymis may be absent or imperfectly formed Such abnormalities result in sterility if bilateral Abnormal relationships between testis and epididymis occur in defects of the urogenital union as previously described

Epididymitis Epididymitis occurs most frequently in young men who have urethritis and aged men who have infection which has followed instrumentation during the course of prostatism

Epididymitis is classified as follows (1) gonorrheal epididymitis (2) metastatic or septicemic epididymitis of which tuberculous epididymitis is an example (3) epididymitis following instrumentation and (4) epididymitis from trauma physical and perhaps sexual excesses

Gonorrheal epididymitis is acute or if mild at the onset tends to become acute later The least trauma excites to an acute relapse This relapsing tendency is short lived The relapses of epididymitis after intervals of months or years are not due to the gonococcus Suppuration in gonococcal epididymitis is common

Metastatic epididymitis arises from distant infections as for instance infection of the upper air passages particularly tuberculosis or an acute pyelonephritis or some other source Epididymitis is usually due to *Escherichia coli* or to *Staphylococcus aureus* The bacillary infection shows the same tendency to gross suppuration to involvement of the testis and cord and to relapse as the epididymitis of urinary disease Staphylococcal infections show a tendency toward healing and less tendency to suppuration and to relapse unless originating from a septicemia

Testicular injury and an ensuing epididymitis may follow trauma to the testicle such as may be sustained from falling astride a fence or from injury incurred during horseback riding The symptoms and course depend on the severity of the trauma Rarely is the testicle injured sufficiently to require its surgical removal

Epididymitis from *sexual strain* the colored man s strain is usually of gonorrheal origin

SYMPTOMS Often the inflammation begins slowly but later flares into an acute attack In still others it remains mild and localized to the distal portion of the epididymis

The acutely inflamed epididymis swells within a few hours to several times its normal size The organ is exquisitely painful and tender the pain extending to the sacrum and to the loin The temperature rises to 103 to 104 F (39 4 to 40 C) In tense symptoms may continue for days until relieved by infiltration of the sheath after which the suppuration burrows through the scrotal fasciae reaches the skin bursts and leaves a chronic fistula

EXAMINATION The acutely inflamed epididymis is a swollen edematous ridge surrounding the normal testis The epididymis may become a large tender mass There may be epididymitis periepididymitis and deferentitis The deferential lesions may extend the whole length of the vas and may be most marked at the lower end of the vas or at the external inguinal ring

DIAGNOSIS The physical findings and history are usually diagnostically definite Epididymitis is distinguishable from torsion by the fact that elevation eases the pain of epididymitis and accentuates that of torsion When a swollen epididymis is associated with prostatitis the condition is usually epididymitis Tuberculous epididymitis can ordinarily be distinguished by the history and the associated lesions and by its slow chronic course and perhaps by suppuration However the diagnosis often is not made until histologic study has been done After any form of epididymitis the prospect of sterility (on the affected side) is great

Sporotrichosis of the Epididymis Sporotrichosis of the testis and epididymis may occur in the course of a cutaneous sporotrichosis in which event the diagnosis

trarectal (seminal vesicular carcinoma) and rectal carcinoma is the mobility of the rectal mucosa over the mass

Diagnosis of primary carcinoma of the seminal vesicle can be established by biopsy and histologic study

THE PROSTATE GLAND

The normal prostate gland is 1¼ to 1½ inches (3 2 to 3 8 cm) wide 1 to 1¼ inches (2 5 to 3 2 cm) long and 1¼ inches (3 2 cm) thick A central depression on its rectal surface separates it into lateral lobes The prostatic tissue is continued uninterruptedly across the median line in the form of a commissure Enlargement of this part constitutes what appears to be a median lobe

Congenital anomalies of the prostate gland are extraordinarily rare and probably not recognizable clinically unless gross anomalies exist

Nocturnal Emissions Nocturnal emissions usually associated with dreams occur in boys and in men Such an emission is simply the discharge of semen while asleep

The fears that boys and young men can build on this physical phenomenon may be excessive The experienced adviser will not even suggest posterior urethroscopy to the patient who complains of only infrequent emissions until a thorough understanding is had of the psychologic aspects of the patient and an attempt has been made to establish an alleviation of fear and anxiety by exercise cold baths and diversion in a normal and unmorbid society to diminish the number of the emissions and to minimize their importance in the mind of the sufferer If these measures fail it may be necessary to have recourse to more formal psychic investigation and then to examine the posterior urethra

Nocturnal emissions may continue in aged men long after they have become impotent Often it must be explained to these aging individuals that the occurrence of these emissions is a normal phenomenon and not the sign of latent virility

Hemospermia (Bloody Ejaculatory Fluid) Hemospermia may be symptomatic or idiopathic in origin

Symptomatic hemospermia may arise from prostatitis or vesiculitis prostatic concretions tuberculosis or carcinoma or may be due to sexual excesses Hemospermia and hematuria may occur simultaneously Symptomatic hemospermia is of no great interest for this symptom alone cannot be evaluated and is thus termed idiopathic hemospermia

Idiopathic hemospermia is not accompanied by hematuria or by pain It is not associated with pus or with sexual strain Massage of the seminal vesicles will often but not always deliver a bloody fluid The amount of blood in the semen varies The semen may appear as pure blood There may or may not be infiltrations palpable about the seminal vesicles The condition seems to occur in middle aged men otherwise in good health The hemospermia may continue for many months or years and shows a great tendency to recur Its sole importance is often related to the apprehension produced by its presence

Prostatorrhea Prostatorrhea occurs when the muscles of the prostate are relaxed so that movement of the bowels squeezes some drops of opalescent sticky fluid out of the meatus This is harmless

Acute Prostatitis Acute prostatitis is probably always associated with varying degrees of vesiculitis and urethritis In young and middle aged men acute prostatitis is often due to gonorrhea acute infections of the upper part of the respiratory tract pyelonephritis and urethral stricture and prostatic massage In old men the causes are pyelonephritis prostatism acute respiratory infections and unnecessary prostatic massage

Urination is frequent painful and urgent A few drops of urine may pass involuntarily every few minutes

4 to 6 inches (10 to 15 cm) in diameter The true nature of this midline pelvic tumor is not known until revealed by surgical removal and histologic study

Concretions and Calculi A number of concretions or small calculi may be present in the vesicles of the aged

Spermatic colic is rare even in those who have calculi It may occur at the moment of ejaculation or during sleep The pain is very sharp and colicky and centralized about an inch up the rectum or at the neck of the bladder or to the testicle A painful and deficient emission may ensue The colic lasts from 10 to 20 minutes and then subsides A hot rectal douche or the introduction of a finger into the rectum with pressure on the offending organ gives relief

Vesiculitis McCarthy and his associates ventured the opinion that vesiculitis is but a corollary to prostatitis However there are three main types of infections of the *seminal vesicles tuberculous gonorrheal and nonspecific Congenital* anomalies such as persistence of the mullerian duct or dilatation or stenosis of the ejaculatory ducts seem to predispose to seminal vesiculitis Congestion or distention over a long period as the result either of prolonged abstinence or of excessive sexual contact also predisposes to development of vesiculitis in the presence of adjacent or focal infection

The commonest form of seminal vesiculitis is produced by extension of infection after posterior urethritis This regional extension which sometimes follows instrumentation of an infected urethra occurs either by migration along the contiguous surface of the mucosa or by lymphatic invasion Infection by means of the blood *stream in the course of systemic disease or from foci of infection is possible,* as is secondary involvement from a focus in the epididymis

Prostatic disease which interferes with proper drainage of the ejaculatory ducts may set up conditions ideal for the development of vesiculitis Urethral strictures which interfere with the proper drainage of urine from the bladder may result in an infection of the urethra bladder prostate and seminal vesicles

SYMPTOMS Dysuria frequency and nocturia may be present and may be the only symptoms A morning urethral drop and a light thin watery discharge at the meatus during the day are often caused by a coexistent inflammation of the prostate and posterior urethra Frequent seminal emissions and early ejaculations are common *Blood ejaculations are pathognomonic of vesicular involvement* Recurrent epididymitis usually indicates chronic infection of the vesicles

A dull pain or ache in the perineum is evidence of congestion due to prostatovesiculitis This is characteristic of true visceral pain which is diffuse poorly localized and frequently of a dull boring nature In the more acute processes the severity increases

Pain in the flanks epigastrium upper abdominal quadrants lower abdominal quadrants and suprapubic region may occur as a result of *infection in the seminal* vesicles prostate and adjacent structures There may also be present aches and pains of the groins buttocks perineum and lower extremities

On digital examination by way of the rectum the swollen and tender vesicle may be felt above and laterally to the prostate

The diagnosis of seminal vesiculitis is made from history physical examination microscopic examination of expressed secretions endoscopy and vesiculography

Carcinoma Carcinoma of the seminal vesicles may resemble on palpation metastatic carcinoma situated on the rectal shelf

The common tumors of the rectal shelf are metastatic lesions and the greatest incidence of occurrence is *found in carcinoma of the stomach It is* frequent that patients who have tumors of the rectal shelf present a history of gastric complaints while persons with primary carcinoma of the seminal vesicles frequently present a history of urinary difficulty and hematuria A point of differentiation between ex

Abscess of the Prostate A prostatic abscess results from localization and resolution of prostatic tissue as a consequence of (1) severe acute or chronic infections of the prostate (2) blood borne infections during the course of severe respiratory infections furunculosis carbuncles and osteomyelitis and (3) instrumentation

There is a sudden onset of acute urinary symptoms such as frequent and difficult micturition Perineal pain is commonly present Fever may reach 103 F or more Leukocytosis of 25 000 cells per cubic millimeter is commonly observed

On rectal examination the prostate is swollen tender and contains a fluctuant mass

Tuberculosis of the Prostate Prostatic tuberculosis is symptomless except for the rigidity which involvement of the gland and of the overlying urethra imparts to the neck of the bladder The resultant achalasia is slight and the symptoms of the tuberculous cystitis are those present in a tuberculous prostate

On examination there may be large and hard masses of periprostatic exudate These masses may also occur about the subacutely inflamed nontuberculous prostate gland

The presence of tubercle bacilli in the vesical urine and the absence of pus and bacilli in the renal urine as obtained by ureteral catheter associated with the physical findings of prostatitis or epididymitis are likely to be symptomatic of tuberculosis Final diagnosis awaits biopsy if this procedure is deemed prudent

Tuberculosis of the Epididymis the Vas and the Testicle Tuberculosis of the external genitalia of the male may begin by focal infection from the blood stream or by extension of the disease from the internal genitalia or by extension to the tunica vaginalis from the hernial sac involved in a tuberculous peritonitis Suppuration may occur at or below the external inguinal ring but scarcely ever above

The acute onset is characterized by swelling fever leukocytosis and pain the chronic onset initially by a lump which may be only slightly sensitive The lesion may remain for years as a nodule of irregular shape without any tendency to suppuration Such nodules are often felt throughout the epididymis

In diagnosis tuberculous epididymitis is to be differentiated from simple epididymitis syphilis calcified hydrocele and neoplasm

In establishing the diagnosis the following clinical features of tuberculosis of the epididymis are important the little round nodules the diffuse infiltration of the epididymis the acute epididymo orchitis the frequency of hydrocele and abscess and especially the frequent sensitiveness to pressure and the involvement of the vas

The discovery of the tubercle bacillus in the urine in the pus massaged from the prostate or in the contents of hydrocele fluid or abscess makes the diagnosis fairly certain

In infancy and childhood the accepted treatment is castration In adult life the tendency toward conservatism is good practice

Prostatic Stones (Concretions) Prostatic concretions occur in men more than 40 years of age These concretions are small round black or brownish bodies impregnated with lime They are situated in dilated glandular acini of the lateral lobes No symptoms are attributable to these concretions Prostatic concretions are diagnosed by the shadows cast on the roentgenogram They are rarely distinctly enough palpated to be thus diagnosed

Prostatism (Stenosis of the Bladder Neck Hypertrophy of the Prostate) The syndrome of prostatism is produced either by narrowing of the neck of the bladder or by the presence of hypertrophy of the prostate in the neighborhood of the urethra Stenosis of the vesical neck and an enlarged prostate may coexist However sclerosis of the vesical neck differs essentially from hypertrophy of the prostate in that it may occur at any age

The pain is situated on the undersurface of the urethra just behind the glans penis in the perineal region, down the thighs in the rectum over the sacrum and often above the pubes

An acutely inflamed prostate gland may impede the outflow of urine A fever the temperature being 101 to 102 F (38 3 to 38 8 C), and leukocytosis are usually present

The prostate is swollen tense and tender The examination is performed gently in order to avoid injury and spread of the infection to the periprostatic tissue If the periprostatic tissue is already inflamed the outline of the gland is completely lost The seminal vesicles may be palpable

The diagnosis is evident from the symptoms and the examination There may be pre existing pyelonephritis or such may develop The inflammation in the vesicles often extends to one or both epididymides

Chronic Prostatitis Chronic prostatitis may follow acute prostatitis or its onset may be insidious

PATHOLOGY Periacinous infiltration and dilatation of the acini are the essential lesion of chronic prostatitis There usually are regions of normal tissue and in some instances the diseased tissue is confined to that part of the gland adjacent to the urethra

The normal prostatic fluid is albuminous in appearance and has a characteristic odor It is alkaline to litmus and acid to phenolphthalein It contains gelatinous transparent lumps of vesicular secretions translucent lecithin bodies columnar and round epithelial cells fewer than 5 leukocytes to the high power microscopic field a few corpora amylacea and often spermatozoa Erythrocytes are present in the secretion obtained by massage of the prostate

Purulent prostatic secretion is thin often yellowish and nonalbuminous in appear ance The intensity of the infection is measured by the number of leukocytes and often by the number of bacteria present

SYMPTOMS Generally a chronic prostatitis is symptomless When symptoms do occur there is pain usually in the perineum or occasionally in the lower part of the back The patient may describe it as a discomfort along the urethra a burning or a desire to urinate Frequent and difficult urination is due to the chronic posterior urethritis or to sclerosis of the vesical neck

A disturbance of the sexual function if present is more likely due to age than to the disease Arthritis and other metastatic infections are rarely if ever due to inflammation of the prostate

EXAMINATION The chronically inflamed prostate gland may appear normal on palpation Usually however the gland is enlarged The enlargement consists of a general softness of the lateral lobes At times the palpating finger may feel nodules of a somewhat increased resistance in the lateral lobes of the gland Synchronously with the examination of a swollen prostate there may be an outpouring of prostatic secretions from the meatus

Prostatic sclerosis is the result of chronic prostatitis of long duration Scar tissue replaces the parenchymatous exudate while infection continues in the acini The scar renders the prostate rigid to palpation and invades the neck of the bladder

Vesiculitis is often a part of the infection which constitutes chronic prostatitis This may or may not result in palpable enlargement In longstanding instances the vesicle may be changed to an irregular cavity containing pus

DIAGNOSIS The diagnosis of chronic prostatitis is made by the discovery of pus and often bacteria in the prostatic secretion Rectal palpation of the chronically in flamed prostate gland may reveal no abnormalities In some patients the prostate may be tense, firm and bulging or soft and boggy

The prostate gland once severely inflamed may not become wholly normal The boggy lobes sometimes but not always can be massaged free of pus but they remain soft

or to the symptoms The symptoms of prostatism are associated with the retention of urine urinary infection and hemorrhage

Theories of the etiology of prostatic hypertrophy are enumerated thus (1) benign epithelial hyperplasia a product of the disturbed sexual physiology of senility which is presided over by the glands of internal secretion (2) arteriosclerosis (3) adenomatous and comparable to cystic diseases of the breast (4) myomatous and comparable to fibromyoma of the uterus and (5) postgonorrheal resulting from stricture of the prostatic ducts

PATHOLOGY There is distention of the glandular acini with proliferation of the epithelium resulting in the formation of masses of glandular tissue The glands are surrounded by varying degrees of hypertrophied muscle and hyperplastic fibrous tissue

SYMPTOMS An important symptom is nocturnal frequency of urination Before there is infection or retention of urine in the bladder the patient has to arise several times at night to urinate This nocturia begins by the patient being awakened with the internal urge to urinate In the beginning he is awakened during the early morning hours as the symptoms increase urination will be regularly spaced through the night An obstruction of the flow of urine is evidenced by its delay in starting its coming in a small stream and dribbling at the end of urination When there is residual urine in the bladder or when the amount of residual urine increases or infection supervenes urination becomes painful as well as difficult Voluntary retention of urine or exposure to cold and wet may bring on acute and complete retention of urine as the first symptom Acute complete retention of urine is the first symptom of importance in almost half the cases Loss of urine may be associated with may precede or follow acute retention

True incontinence of urine does not occur as the result of prostatism The loss of urine that does occur is overflow from the atonic distended bladder The overflow of urine is the result of an irritation of the bladder the loss of urine is involuntary

Hematuria is common from prostatism and the presence of blood in urine is not necessarily indicative of the presence of malignant disease The bleeding is usually due to an ulceration of the middle lobe to acute inflammation or perhaps to stone in the bladder Bleeding unavoidably occurs from the passage of instruments

Debility and age are the potent forces which annul the sexual powers Such a loss of function is not due to prostatism The congested prostate may excite abnormal erections which may be interpreted by an atherosclerotic brain as the return of youth and vigor which may lead the patient to the altar of marriage and soon thereafter to disaster

In some aging men an uninfected bladder may reach chronic distention with back pressure extending upward and involving the kidneys before there is known serious trouble Under such circumstances the first symptom recognized is a loss of weight and strength accompanied with constipation and dry mouth This evidence may be considered trivial until fever appears

Inflammatory complications occur sooner or later in every case and are often initiated by catheterization The cystitis often follows even necessary catheterization The acute cystitis which follows the passage of a catheter begins acutely often with a chill Spontaneous cystitis is often mild and superficial and may not cause pain or frequency of urination Spontaneous infection rarely if ever occurs before the use of the catheter or massage of the prostate for the chronic prostatitis which is present in every instance of infection

Epididymitis may occur spontaneously or may follow instrumentation Often epididymitis is a symptomless chronic induration at one end of the epididymis Epididymitis may be severe however and suppuration occasionally supervenes

Seminal vesiculitis is common and usually is clinically unimportant

The course of the disease is often interrupted by an acute renal infection due to vesicoureterorenal reflux of urine The symptoms of the vesicoureterorenal reflux

Stenosis of the Neck of the Bladder Stenosis of the vesical neck often is *congenital* It may become manifest at any age In infancy the bladder is so vigorous that it dilates with little sacculation in comparison to the megaloureter that ensues In early life and up to 50 years of age the result of sclerotic obstruction is often diverticularization of the bladder and somewhat less dilatation of the ureter

Stenosis whether in the prostate or in the vesical neck cannot be dismissed with the statement that it is due to inflammation for gonorrheal urethritis and prostatitis are rarely followed by sclerosis sufficient to obstruct the neck of the bladder Prostatic abscess rarely results in sclerosis

Stenosis is commonest between the ages of 30 and 50 years 20 years or so *before the age of maximal incidence of prostatic hypertrophy*

The lesion of the vesical neck comprises chronic inflammation and sclerosis which involve the mucosa the underlying tissues and the sphincter often a secondary hypertrophy of the musculature of the bladder follows The result of such sclerosis is achalasia rigidity of the vesical neck

SYMPTOMS In most instances sclerosis of the neck of the bladder does not cause significant symptoms Every severe posterior urethritis leaves some scarring about the vesical neck These scars are not clinically sclerosis until there is retention of urine The stream loses much of its force the residuum is constantly at least 50 ml The urinary stream is slow and small difficult to start and ends with the dribbling of urine There is nocturia

EXAMINATION Routine examination of the abdomen of a debilitated child may reveal a distended palpable bladder which indicates the need for a urologic investigation

Young men affected by stenosis of the neck of the bladder often have been embarrassed when they were boys by being unable to participate in micturition contests and like girls by having to urinate in the squatting position

When the age is attained in which the prostate gland may become hypertrophied if there is any sclerosis of the vesical neck symptoms will be more intense and will occur earlier The symptoms of sclerosis may be overshadowed if there is considerable general hypertrophy The patient often has achalasia and sclerotic lateral intra urethral lobes which give the symptoms common to prostatism

In *traumatic sclerosis* as the result of external trauma the urethra may be ruptured *for instance as a complication of a fracture of the pelvis* The more familiar form of traumatic sclerosis however is that which follows prostatectomy or resection After prostatic resection the onset of the lesion cannot be predicted It may appear causing retention of urine before the patient has left the hospital Symptoms rarely occur however before several years have passed

DIAGNOSIS The sclerotic bladder on endoscopic examination is almost always inflamed and edematous cystic or ulcerated Only the well trained urologist can distinguish the constricting effect of achalasia from the spasm of inflammation and *estimate its degree Also an expert and only an expert* by cystometric examination can distinguish the neurogenic atony such as is observed in tabes dorsalis from the muscular atony of the bladder due to sclerosis

Uncomplicated sclerosis of the neck of the bladder is suspected whenever retention of urine in the bladder is found and rectal touch reveals no enlargement or irregularities in the prostate A small residuum of urine in the bladder of a patient who has arteriosclerosis and who is neurotic or psychotic does not interfere with the force of the urinary stream

Hypertrophy of the Prostate There may be a general hypertrophy of the prostate without significant obstruction to the flow of urine The syndrome of prostatism depends on either the degree of sclerosis of the neck of the bladder or the presence of hypertrophy of the prostate in the neighborhood of the urethra The amount of residual urine bears no essential relation to the size of the hypertrophy

partial or complete paralysis of the bladder The absent ankle jerk is common enough in old age yet when this sign is noted accompanied with the absence of deep muscle sense the spinal fluid should be examined

The prognosis of prostatic retention depends on the patient s age and on the character of the prostatic enlargement

Tumors of the Prostate Gland ***Benign Tumors*** Occasionally *fibromas* and *adenomas* are discovered in the prostate These usually present the symptoms of benign hypertrophy of the gland

Cysts of the prostate are usually single and apparently due to occlusion of one of the ducts The cyst may project toward the urethra or the bladder or from the posterior surface or upper border of the gland Intra urethral cysts grow large enough to cause complete retention of urine Retrovesical prostatic cysts grow to enormous size but do not cause symptoms except from their size

The intra urethral cysts usually project characteristically from the upper portion of a lateral lobe Their translucency may be appreciated on cystoscopic examination

Malignant Tumors CARCINOMA Cancer of the prostate is present in 1 of every 5 patients who have prostatism Prostatic cancer arises from the epithelium in the terminal acini of the ducts as a single focus or as multiple foci The disease may remain confined to the gland for a long time Finally however the basement mem brane is perforated and the neoplastic process passes into the lymphatic structures

The structural type of prostatic carcinoma varies widely The highly malignant tumors are nearly all highly anaplastic They arise in prostates which have not been the seat of prior disease The tumors are small regular in outline and elastic The lymphatics are invaded uniformly and the small veins are thrombosed early by tumor There is immediate and widespread metastasis to the bones which may be the first evidence of the disease

The more definite adenocarcinomas are less highly malignant than those charac terized by a high degree of anaplasia They invade the stroma more slowly but pro duce small very hard irregular tumors The lymphatics in the organ are invaded and metastasis to the regional lymph nodes occurs early in the disease There is a high incidence of perineal sacral and sciatic pain Some adenocarcinomas may arise in large glands which are often of soft texture Demonstrable metastasis is rare The true nature of the process is often not discovered until histologic study is made

Metastasis of carcinoma of the prostate occurs mainly through the lymph vessels The lymph vessels of the prostate are distributed with the internal iliac vessels and drain to the lumbar lymph nodes which are continuous with the mediastinal lymph vessels and nodes In the lumbar region the nodes are in close apposition to the vertebrae so that metastasis to the vertebrae is likely a retrograde extension In the highly malignant tumors the left supraclavicular lymph node is enlarged

Direct extension of the cancerous process into the perineural lymphatics in the prostate gland involves the sciatic plexus and gives rise to sciatic pain Bilateral sciatica in middle aged men often suggests cancer of the prostate Metastasis less frequently involves the parenchyma of the lungs or liver The process in bone is at first destructive later there is osteoplasia The deposition of calcium at the periphery of the tumorous deposit gives rise to the roentgenologic findings described as osteo plastic metastasis In some cases the osseous process is wholly destructive and remains so

The initial *symptoms* of cancer of the prostate comprise the urinary symptoms and pain The common urinary symptoms are frequency and difficulty of urination Other symptoms associated with these are nocturia hematuria urgency and incon tinence Pain accompanying urination backache sciatica or pain in the perineum rectum or penis may be the cause for the patient to consult a physician

Occasionally and usually when the tumor is highly malignant urinary symptoms do not occur even in well advanced cancer of the prostate When present the

are the same as those of the urethral chill but of longer duration The renal infection usually becomes chronic Pyelonephritis may not be present but if present it may remain slight for years or may soon be complicated by hydronephrosis or may de velop into pyonephrosis

An occasional patient who has prostatism may have a stone in the bladder Stones may be present in those who have but little retention If removed stone will recur repeatedly until the prostate is resected

Examination in Prostatism A general physical examination is essential for dis closing the effects of prostatism and for determining other disease which is often present An oval mass above the pubes palpable bimanually is a distended bladder An irregular mass above the pubes may be a carcinoma of the prostate extending around the bladder an oval mass not palpable bimanually is probably a urachal cyst

The *rectal examination* should not be made until the bladder has been emptied Observe the patient while he urinates Often there will be this sequence of events start—stop—start slow small stream which ends with weak spurts of urine and finally dies away with dribbles and then drops of urine which trickle over the end of the penis and fall in a backward direction

With the patient leaning forward at a right angle the examiner s finger is moved across the *prostate gland* The findings of this examination are described with regard to lobal enlargements smoothness and mobility An enlargement of the lateral lobes is manifested by bulging into the rectum If the lateral lobes are widely separated there may be enlargement intravesically if close together the enlargement may be intra urethral The hypertrophied lobes are usually symmetric, tense and smooth Irregularities felt on or in a lobe may be due to stone inflammation or carcinoma The benign prostate is relatively movable the malignant tends to become fixed owing to the periprostatic extension of the disease

The *contraindications to catheterization* are (1) complete retention with disten tion and (2) acute infection of the prostate and fever Catheterization of a patient who has complete retention with a distended bladder palpable above the pubes is deferred until the patient is in hospital prepared for decompression

As the patient urinates the sluggishness of the stream is noted If there is com plete retention the patient should be sent to hospital for decompression before further examination is made though if the retention is acute the bladder may be emptied at once by catheter If the patient has not been catheterized before a rubber instrument is passed with gentleness for it is this first catheterization that often infects the patient and may result in serious illness and death The amount of residual urine is measured

In the presence of small and even of medium sized enlargements cystoscopic examination is often necessary in order to search for prostatic bar diverticulum and tumor

If the patient is to be operated on and is not to be examined cystoscopically an *excretory pyelogram* is made in order to detect unsuspected urinary stones and func tional or anatomic defects in the kidneys Concentration of urea in the blood is determined and often blood grouping and Rh factors are determined

Diagnosis of Prostatism The diagnosis of prostatism is made by the summa tion of the findings on examination which have been enumerated Nocturnal polyuria of nephritic or of arteriosclerotic origin is often confusing and is differentiated so that a precise diagnosis of prostatic hypertrophy can be made Renal insufficiency is not the cause of these symptoms if the range in specific gravity of the urine is from 1 010 to 1 020 or more

Tabes dorsalis cannot be distinguished either by the serologic reactions or by absence of knee jerk alone The Argyll Robertson pupil is often diagnostically helpful The absence of ankle jerk in association with a loss of deep muscular sense may be the only outward sign of tabes that is early yet sufficiently ad cause

It is realized that regions of increased circumscribed hardness situated within the prostate gland are not pathognomonic of cancer Such nodules may be due to stone chronic disease tuberculosis or chronic prostatitis

SARCOMA Sarcoma of the prostate is a highly malignant tumor of connective tissue origin which usually occurs only in infants and children its rate of growth is extremely rapid it metastasizes readily to the lungs and is frequently fatal within a few months of its onset The symptoms which it produces are purely obstructive complete retention following rapidly the increased difficulty in urination Rectal examination reveals the presence of the large elastic mass which has replaced the prostate Up to date sarcoma of the prostate has been uniformly fatal and the only available treatment is palliative

THE PELVIS

The male pelvis is fashioned pre eminently for locomotion it is both heavier and rougher than the female pelvis The female pelvis in addition to the functions of locomotion has the added function of gestation The cavity of the female pelvis is larger and shallower than that of the male pelvis It is smoother its bony prominences are less conspicuous The symphysis pubis is narrower and the sacrum is shorter and less curved The acetabula are set wider apart

Pelvic Hernias Hernial protrusions of the pelvic contents may occur through the upper portion of the obturator membrane following the vessels and nerve Such a protrusion is called an obturator hernia It makes its appearance in Scarpa s triangle and is covered by the pectineus muscle Sciatic hernia is the name given to those forms in which the intestine escapes through the great sciatic notch passing just above or just below the pyriformis muscle Perineal hernias may push down between the rectum and bladder and bulge in the perineum They may pass between the coccygeus and levator ani muscles or between the fibers of the latter and bulge into the ischiorectal fossa or forward into the labium of the female

Pain in the External Genitalia The *labia majora and minora clitoris vestibule and urethral meatus* are sensitive to heat cold and touch The skin of the labia majora is more sensitive than the skin or the mucous membrane of the labia minora to heat cold touch or pinching

The *urethras* of both men and women before instrumentation are usually sensitive whereas a much instrumented urethra in either sex may become insensitive Gentleness persuasion and reassurance to induce relaxation before and during instrumentation may overcome much of the apparent sensitiveness The pain elicited by instrumentation of the urethra is due in part to the sudden stretching of a contracted muscle rather than to the sensitivity of the mucous membrane The passage of the cystoscope may elicit a desire to defecate

Overdistention of the bladder with solutions at any safe temperature produces first a feeling of fullness and then of pain in the suprapubic region and in the urethra associated with an intense desire to urinate The pain from overdistention of the bladder in men is referred to the end of the penis In women the pain from overdistention is referred to the labia minora and clitoris

Pain from stimulation of a *ureter* may be felt ipsilaterally and its location in the skin follows approximately a line drawn along the lateral edge of the rectus muscle The area of the cutaneous site of pain may be hyperalgesic as tested by pin prick

When the *ureterovesical junction* and the first 2 inches (5 cm) of the ureter are distended or stimulated pain may be felt in the suprapubic region along the urethra and is accompanied by an intense desire to urinate In some instances the pain extends from the suprapubic region up the midline of the abdomen as far as the umbilicus and may spread across the abdomen In an occasional individual stimulation of the ureter may cause pain along the anterior surface of the thigh and leg to the knee

Dilatation of the normal *renal pelvis* always elicits pain that is felt in the costovertebral angle There is no recognition of heat or cold sensation in either the renal pelvis

symptoms of urinary difficulties and pain become more severe as the disease progresses

On *rectal examination* there may be an irregularity of the contour of the prostate or differences in density in various parts of the gland In advanced prostatic cancer there is an extension of the hard irregular infiltration beyond the confines of the organ about the base of the bladder and the seminal vesicles fixing the organ in the pelvis If there is a large and hard single nodule confined to the organ it may be due to the presence of stone Multiple small calculi may produce crepitus when pressure is brought to bear by the examining finger

Cystoscopic and endoscopic examinations are of little value in early cancer of the prostate The residual urine is measured and this too is variable Roentgenologic examination of the bony pelvis and lower lumbar vertebrae will often reveal the presence or absence of metastasis to the bone or the presence of stone in the prostate

The changes in the values of the acid and alkaline phosphatase in the blood may be of great aid in detecting metastatic lesions in the bone not yet visible by roentgenologic examination

Acid phosphatase is an enzyme which is found in the epithelial cells of the prostate gland in sexually active men and in the cells of prostatic cancer Values of more than 10 Bodansky units per 100 ml of serum are significant Normally there is a small amount (less than 3 Bodansky units) of this substance in the blood serum The enzyme owes its existence to the sustaining influence of the male sex hormone After castration it disappears from the prostatic epithelium and a coincidental shrinkage of the tissues ensues

It is well established that sustained elevation in the serum acid phosphatase level occurs when malignant metastasis has involved the bone marrow and lymph nodes Investigations have shown that transient elevations in the serum titer of this enzyme may be detected after the trauma of operation on the prostate gland and subsequent to digital manipulation but not as the result of prostatitis benign prostatic hypertrophy or tumor metastatic to the prostate from other organic structures Thus sustained abnormal elevations of this enzyme are pathognomonic of metastatic prostatic carcinoma

Alkaline phosphatase is produced by bone as an essential agent for growth and repair The normal values are 1 5 to 4 Bodansky units per 100 ml of serum in adults and 5 to 14 units per 100 ml of serum in children With few exceptions its serum level reflects bone activity in the absence of hepatic disease However in the presence of known metastatic prostatic carcinoma its elevation may signify osteoblastic response to invading tumor and thus serves as an index of the extent of such activity When elevations are present in cancer of the prostate gland the serum level of the alkaline phosphatase is generally higher than that of the acid phosphatase After castration when remission occurs the acid phosphatase level promptly drops to normal whereas there is a temporary rise in the alkaline phosphatase value followed later by a drop toward the normal level This behavior affords an index of the effectiveness of treatment

The presence of an abnormal serum acid phosphatase level in the absence of evident metastasis in a patient who has prostatic cancer implies a poor prognosis The presence of a normal serum acid phosphatase value in a patient who has metastatic prostatic carcinoma does not necessarily imply poor response to endocrine therapy Serum alkaline phosphatase concentration is not in itself specific for prostatic carcinomatous activity without metastasis to the bone

The *diagnosis* of cancer of the prostate is never certain until biopsy is made at the time of operation In the presence of urinary difficulties metastasis to bone and sciatic or perineal pain suggest the diagnosis of cancer of the prostate Add to this a hard fixed prostate gland on rectal examination and the diagnosis is almost conclusive

Primary carcinoma of the prostate must be differentiated from secondary carcinoma metastatic from some other primary lesion particularly rectal carcinoma and from sarcoma prostatic cyst and tumors of the rectum sigmoid and base of the bladder as well as from the more usual prostatic lesions

broad ligament Cysts arising from Gartner s duct are sometimes found in the vagina In the male cysts arising from the wolffian duct are (1) encysted hydrocele of the testicle and (2) general cystic disease of the testicle Cysts arising from the persistence in the male of Muller s duct have also been observed in the prostate and seminal vesicles but they are exceedingly rare

This discussion is continued with the description of the disease of each organ

THE EXTERNAL FEMALE GENITALIA

The labia majora meet anteriorly in the anterior commissure and posteriorly in the posterior commissure They vary greatly in size and in color often the median sides of the labia are very much more pigmented than the surrounding skin The space between the posterior commissure and the anus about 1¼ inches (3 2 cm) is the perineum

The labia minora divide anteriorly to form the prepuce above the clitoris and the frenum on its lower surface Posteriorly they decrease in size to a thin crescentic fold of mucous membrane called the fourchet Like the labia majora the labia minora vary greatly in size and particularly in the anteroposterior width They often project outside of the labia majora The space between the labia minora is the vestibule The meatus or urethra is in the vestibule 1 inch (2 5 cm) behind the clitoris It is surrounded by a ring of mucous membrane and when a catheter is to be introduced the urethra can be seen by separating the labia minora or it can be recognized by the sense of touch The openings of the para urethral ducts are just below and to the outside of the meatus The vulvovaginal glands (of Bartholin) empty on the inner side of the labia minora in the sulci between them and the hymen

The hymen partly occludes the lower end of the vagina across its posterior portion The carunculae hymenales are the remains of the ruptured hymen The fossa naviculars is the space between the hymen and the fourchet

THE VULVA

The vulva includes the mons veneris labia majora labia minora clitoris and prepuce vestibule hymen urethral meatus and Bartholin s and Skene s glands Variations in size and true hypertrophies of the clitoris and labia minora are common In rare instances there may be a complete double vulva

The vulva becomes edematous when there is edema of the extremities as in congestive heart failure Varicosities of the vulva may be associated with varicose veins of the legs Hematomas occur in the normal vulva after trauma They may become large and painful

Acute Vulvitis Acute nonvenereal vulvitis includes intertrigo a diffuse reddening of the skin of the vulva and folds of the thigh which occurs in infants and obese women mainly during hot weather Follicular vulvitis or hair follicle infection is common Furunculosis or boils may develop in hair follicles Noma a gangrenous vulvitis in young girls in tropical climates follows severe systemic infections such as measles scarlatina diphtheria typhoid fever smallpox and diabetes The diabetic vulvitis is often of fungous origin and is usually accompanied by vaginitis

Acute Venereal Vulvitis *Gonorrhea* The vulva except for Bartholin s and Skene s glands is not ordinarily involved in gonococcal infections and the majority of the infections of Bartholin s gland are not due to gonorrhea

Vulvitis with redness and swelling is rare in the adult woman It is occasionally seen in acute gonorrhea associated with pregnancy Vulvovaginitis is common in children The vagina is inflamed and there may be redness and swelling of the vulva

Weaver stresses the value of bacteriologic cultures in the detection of the gonococcus in contrast to the inadequacy of the stained smear pointing out that in known

or the ureters Lennander observed that the outer surface of the kidney could be pinched cut or have heat and cold applied without eliciting any sensation

Distention of the right renal pelvis may produce pain extending down toward the right groin and sometimes spreading to the urethra and testicle Ockerblad and Carlson observed that faradic stimulation *within the renal pelvis* always elicited pain which was felt on the back at the costovertebral angle and that the pain never extended around to the front nor upward from this point

THE FEMALE GENITALIA

Aberrations of the Female Genitalia and Their Correlations with Homologous Structures in the Male The wolffian duct with a collection of tubules is known as the wolffian body and reaches its full development in the seventh week On one side of the wolffian body develops the sexual gland which later becomes in the male a testicle in the female an ovary By the end of the second month the kidney develops at the caudate extremity of the wolffian body At this time the bladder is connected by the urachus with the stalk of the allantois The lower end of the bladder is connected with the extremity of the intestinal tract through a dilatation called the urogenital sinus The union of the urogenital sinus and intestine forms the cloaca At the time the wolffian body is developing there appears alongside of it a tube called Muller s duct This duct atrophies in the male but in the female becomes the uterine tube uterus and vagina The ureter is developed and becomes connected with the lower portion of the bladder

The wolffian duct and the mullerian duct until about the third month empty into the urogenital sinus Differentiation of the sexes begins about the third month and is well advanced in the fifth As stated the sexual gland in the male becomes the testicle and passing from its lower end is seen the gubernaculum In the female the sexual gland becomes the ovary and the round ligament passes from its lower end The wolffian body after performing temporarily the functions of a kidney disappears leaving some times a small cyst known as the hydatid of Morgagni (stalked hydatid) attached to the upper part of the epididymis in the male and in the broad ligament near the ovary in the female In the lower portion of the hydatid are the remains of the wolffian body comprising some short closed tubes in the tail of the epididymis known as the para didymis or organ of Giraldes in the male and the paroophoron of the broad ligament in the female The wolffian duct which forms the vas deferens and part of the epididy mis in the male forms the atrophied paroophoron in the female to the inner side of the ovary

The parovarium or organ of Rosenmuller is the remains of the middle set of wolffian tubules and in the male forms the epididymis In the female it is almost always present as a horizontal tube with shorter tubes connected with it between the layers of the broad ligament near the ovary The wolffian duct may persist as a small tube known as the duct of Gartner in the broad ligament close to the uterus and vagina In the male the mullerian ducts atrophy and form the sinus pocularis of the prostate Part of them may persist patulous as Rathke s duct As stated in the female they form the uterine tubes uterus and vagina

Exstrophy of the bladder epispadias hypospadias and various forms of her maphroditism result when the walls of the bladder and urethra and external genitals fail to develop in the median line Should the urachus not close a fistulous tract leads from the bladder to the umbilicus from which urine discharges Cysts may also form in its course If the partition between the rectum and the anus is not ab sorbed there is formed one of the varieties of imperforate anus In some cases the rectum empties into the urethra or bladder thus forming a cloaca Should the testicle become arrested in its descent from the region of the kidney it forms what is known as undescended testicle It may be arrested within the abdominal cavity in the inguinal canal or near the external abdominal ring

The paroophoron gives rise to cysts which have a tendency to develop between the layers of the broad ligament and are papillomatous inside The parovarium also gives rise to cysts which likewise tend to burrow between the layers of the

The lesion may occur on other portions of the skin such as the face neck and back Often the gross appearance is most characteristic

The diagnosis is based on the results of examination of scrapings from the ulcer stained with Wright s stain or Giemsa s stain the presence of Donovan bodies (Donovania granulomatis) as clusters of cocci some occurring particularly in large endothelial cells but also in monocytes leukocytes and in clusters outside of cells partially contained in a membrane has been accepted as diagnostic Some of the organisms are surrounded by thick dark capsules All other venereal diseases must be ruled out by serologic examinations and skin tests

Lymphopathia Venereum and Psittacosis Viruses Synonyms for lympho pathia venereum are climatic bubo tropical bubo venereal bubo fourth venereal disease sixth venereal disease lymphogranuloma inguinale granulomatous lympho matosis poradenitis esthiomene and maladie de Nicolas et Favre

Lymphopathia venereum is a protean disease usually transmitted by venereal contact and manifested by both constitutional symptoms and acute and chronic tissue changes in the inguinal and anorectal regions It is caused by a virus which is similar to that which produces psittacosis

In the first stage it forms a superficial ulcer which varies from a few millimeters to $^4/_5$ inch (2 cm) in diameter This primary lesion may occur at any point in the vestibule or inner surfaces of the labia commonly on the urethra or in the fourchet It may disappear soon or may develop into a deep lesion on the vulva or on the sides of the vaginal orifice completely destroying the urethra If the primary lesion occurs on the upper two thirds of the vulva the lymphatics drain to the inguinal lymph nodes with formation of bubo and perhaps multiple draining sinuses Not infrequently the lymphatic channels of the vulva are blocked forming elephantiasis chronic hypertrophic vulvitis or esthiomene Inguinal involvement in women is less common and less severe than in men If the virus involves the urethra ulceration and strictures may follow Because of the lymphatic drainage from the primary lesion rectal strictures are commoner in women than in men Diagnosis of the late lesions may be fairly accurate from the gross appearance but should be substantiated by exclusion of other lesions

Lymphopathia venereum must be differentiated from chancroid bubo due to pyogenic lesions of the lower extremities tuberculosis of the inguinal lymph nodes gonorrhea syphilis granuloma inguinale balanitis plague tularemia carcinoma and tuberculosis of the rectum and ulcerative colitis Films of pus or biopsy ma terial are examined for elementary bodies by means of the Macchiavello stain Sus pected material should be inoculated intracerebrally into mice The serologic tests for syphilis Ducrey s skin test darkfield smears scrapings and biopsy and the Frei test all are performed as parts of the differential diagnosis

The Frei test consists of the intradermal injection of 0 1 ml of antigen and 0 1 ml of control material Results of the test are read 48 to 96 hours after injection and if the reaction is positive the central induration of the papule caused by the antigen measures 6 by 6 mm or more while that caused by the control material measures 5 by 5 mm or less

Reaction to the Frei test usually becomes positive 7 to 40 days after the onset of the adenitis Negative reactions in definite cases are rare they are usually due to such factors as the menses septicemia fever tuberculosis and coexistent early syphilis or early chancroid

Axelrod tested the yolk sac virus antigen of venereal lymphogranuloma and found that the results of the virus antigen are consistent with positive history and signs of the disease This test has not come into wide clinical use Bedson and co workers demon strated that titers of 1 32 or more in the complement fixation test made with steamed virus of lymphogranuloma venereum and the serum samples from patients in whom the clinical observations are compatible with a diagnosis of lymphogranuloma venereum

cases of gonorrhea in women gram negative diplococci may be found in stained smears in less than 20 per cent of the cases whereas cultures on proteose peptone hemoglobin agar yield a correct diagnosis in more than 75 per cent At Weaver s laboratory when oxidase positive gram negative diplococci are found on cultures they are further studied from the standpoint of their fermentation reactions on various sugars The gonococci will ferment glucose but none of the other sugars *Neisseria sicca* ferments all the sugars except mannitol It is this organism to which Weaver calls special attention because physicians who depend on staining procedures for the diagnosis may make false positive diagnoses

Syphilis A chancre is often situated near the vaginal orifice on a labium majus or minus or about the rectum It is a grayish granular ulcer round or oval from ⅖ to ⅘ inch (1 to 2 cm) in diameter raised flat and firm Multiple chancres occur in the folds where the labia are in contact The diagnosis is proved by darkfield examination demonstrating the spirochetes

Secondary lesions occur as condylomata lata and mucous patches The tertiary lesion forms deep crater like ulcerations Diagnosis is suggested by results of the histologic examination and disappearance of the lesion under antisyphilitic therapy Positive results of serologic tests are present when the lesions are secondary and tertiary but by no means are these diagnostic of vulval syphilis Tertiary syphilitic manifestations originate extremely infrequently in the vagina The isolated submucous gumma breaks down early and appears in the form of a characteristic ulcer

Granuloma Inguinale There are three infections of the anorectal regions and external genitalia of both men and women which are characterized by ulcerations and inguinal lymph node involvement ulceration of the skin or mucous membranes and often abscess formation These are (1) granuloma inguinale (2) lymphopathia venereum and (3) soft chancre (see also page 410)

Granuloma inguinale is a bacterial disease the exact causative bacterium is unknown Lymphopathia venereum is a viral disease and soft chancre is caused by *Hemophilus ducreyi*

Granuloma inguinale originates from the organisms which produce Donovania granulomatis The disease is largely limited to Negroes or to persons of mixed race who have Negro blood Granuloma inguinale or granuloma venereum or pudendal ulcer is primarily a disease of the skin producing an ulceration which has a tendency to extend from the vulva to the inguinal regions

Packer Turner and Dulaney called attention to the fact that the earlier concepts of the progress of this infection solely by serpiginous spread or by auto inoculation have been broadened in recent years as a result of accumulated evidence of lymphatic spread and involvement To lymphatic spread has also been attributed the extensive involvement of the pelvic organs following granuloma inguinale of the cervix and vagina Of significance are the pronounced systemic manifestations which are frequently associated with lesions in this region particularly when complicated by pregnancy These systemic manifestations indicate that granuloma inguinale far from being strictly a local disease of the genitalia has potentialities for widespread dissemination throughout the body either of the specific etiologic agent or of the products of secondary invaders The former possibility has already been demonstrated by a few reported cases of metastatic lesions of bones and joints in distant parts of the body following lesions of the genitalia Thus has evolved from the early beliefs of the limited implications of this disease a clearer understanding of the possibilities for widespread dissemination which the disease possesses

The initial lesion begins as a papule ⅖ to 1⅔ inches (1 to 4 2 cm) in diameter develops into an ulcer and may denude the skin of the vulva perineum and gluteal folds The cutaneous margin of the ulcer is raised and pigmented and has a serpiginous border and a bright pinkish red granular appearing base which reflects light

glands These lesions may itch Urine may be a constant cause of irritation and burning

Collections of smegma round the clitoris or in the folds between the labia are prone to produce irritation and mild itching sensations A persistent and irritated hymen everted vaginal mucosa due to relaxed vaginal outlet gonorrheal warts leukoplakia vulvitis and ulcerative or neoplastic diseases of the vagina or external genitalia all may cause itching

Protective pads or intravaginal tampons used to absorb menstrual and leukorrheal secretions may be responsible for irritations and itching Dermatologic factors include infections of the hair follicles and sweat glands fissures in the skin dry scaly dermatitis in which the skin becomes diffusely irritated reddened purplish or grayish and appearing as if it had been scalded A rare form of pruritus is itching which involves the fossa navicularis vestibule or interior of the vagina

Vaginismus This is a painful spasm of the vagina due to local hyperesthesia Vaginismus is termed superficial when the discomfort is at the entrance of the vagina It is termed deep when the discomfort is situated deep and seemingly in the bulbocavernosus or in the levator ani muscles In some instances the discomfort is apparently due to spasm of the constrictor vaginae muscle Vaginismus is termed psychic when there is extreme aversion to coitus The act is attended apparently by painful contraction of all of the muscles of the vagina

Nymphomania The term nymphomania is employed to designate an exaggerated sexual desire in a woman It is a condition the opposite of vaginismus and like the psychic form of vaginismus it has nothing to do with the state of physical health or the hormones of the glands of internal secretion These women often have had abnormal sexual experiences and frustrations This condition is often present as a part of schizophrenic psychosis

On examination the labia are large and have a rugged appearance and may be excessively pigmented

Leukoplakia Vulvae The term leukoplakia vulvae refers to areas of thickening on the skin The cause of leukoplakia is unknown The disease is characterized by translucent white patches of irregular size which may have indefinite borders or may be widespread It occurs particularly about the clitoris labia minora and on the perineum It may occur inside the rectum Very commonly there is associated kraurosis Pruritus is common and severe Lichen planus and leukoplakia bear a great resemblance to each other when situated on the vulva In lichen planus similar lesions will be present on the flexor aspects of the forearms fronts of the legs and perhaps in the mouth

In the differential diagnosis lichen planus is to be excluded Lichenification is differentiated by the fact that it commonly follows protracted pruritus The spots of lichenification are thickened diffuse usually symmetric situated more on the outer parts of the labia majora and between the buttocks round the anus than elsewhere Lichenification is usually moist and has fissures

Leukoplakia represents a truly premalignant lesion

Tumors of the Vulva ***Benign Tumors*** *Condyloma acuminatum* (verruca acuminata) or venereal wart is often the result of unsanitary conditions Warts of this type are common in the obese during pregnancy and in association with gonorrhea

Verrucae acuminatae occur on the vulva in the vagina and urethra and on the cervix They may be a few millimeters in diameter or a cauliflower mass that almost covers the external genitals They are distinguished from the flat sessile condylomata lata

Fibromas and *fibrolipomas* of the vulva are common The fibroma is hard and movable and is covered by healthy skin It is usually small The fibrolipoma is

suggest active infection with the virus of lymphogranuloma venereum The serums from human infections with the viruses of psittacosis and lymphogranuloma venereum show a high degree of cross reaction in the complement fixation test with antigens made from these two viruses The Frei test also merely indicates that infection with a virus of the lymphogranuloma venereum and psittacosis group has occurred A positive reaction to a Frei test together with a positive reaction to a complement fixation test at a serum dilution of 1 16 or more in a patient suspected of having lymphogranuloma venereum is good evidence of active infection with the disease Acid extracts of lymphogranuloma venereum and psittacosis viruses appear to give specific reactions when injected intradermally in human infections with these two viruses

Herpes Herpes genitalis in women is due to the herpes simplex group of viruses which affect the mouth and nose and the male genitalia Bullous lesions may occur singly or multiply or extensively Ordinarily however a single vesicle followed by an ulcer appears about the labium or the vaginal orifice

The blisters may break and form numerous small superficial tender ulcers At times it has been thought that herpes is transmitted by sexual intercourse but it often occurs without intercourse Occasionally the ulcers recur with each menstrual period In such cases the diagnosis is not difficult Smallpox vaccine has been used for treatment with some success

Ecthyma of the Vulva Ecthyma of the vulva resembles condyloma latum The lesions begin as pustules and form depressed ulcerations *Staphylococcus aureus* and *Streptococcus pyogenes* may be cultured

Kraurosis Vulvae In kraurosis vulvae the skin is white red or mottled and appears dry and thin There is a progressive shrinkage of the vaginal orifice labia minora and clitoris and of the labia majora The mucosal border seems to extend beyond the normal confines and has a smooth glistening unhealthy appearance Kraurosis is common after the menopause Pruritus may be associated with secondary infection incident to injury of the skin from scratching Inflammatory ulcerations are slow in healing mainly because of persistent irritation

Disease of the Vulva Due to Disturbances of Innervation or to Absence of Psychic Control *Pruritus Vulvae* Intense itching is a symptom of various skin diseases and may occur idiopathically as a neurosis Itching may be an undesirable normal impulse for it is present in varying degrees each day in every man woman and child If judgment can be rendered from observations of them scratching or making various rubbing movements as substitutes for scratching all mammals are likewise troubled almost constantly by itching about the anus and genitalia

In women genital and perineal pruritus is common and is perhaps the most disabling form of pruritus It may be temporary and insignificant such as the itching that is often felt during menstruation on the other hand it may become a fixed obsession The irrepressible impulse to scratch creates a vicious cycle for scratching makes itching As an obsession a pruritus gives its victim very little rest or peace and eventually there is present a deterioration of the disposition and health

Senile and atrophic changes in the skin due to a normal or artificial menopause or irradiation therapy are frequently accompanied by pruritus In these conditions the skin becomes thin and wrinkled loses its resistance to infection and becomes susceptible to irritation

Pruritus ani and vulvae are common in obese women whose genitals are constantly wet with perspiration

Rectal discharges and intestinal infections may cause pruritus Also disorders such as hemorrhoids proctitis parasitic intestinal infections fissure and rectal polyps are frequently the basis of the trouble Pruritus may be ascribed to diabetes mellitus However in practice the impression is gained that most instances of pruritus are not associated with diabetes

In some cases pruritus starts as urethritis or as an infection of the periurethral

Urethritis Urethritis is an inflammation of the mucous membrane of the urethra It may be either acute or chronic

ACUTE URETHRITIS The female urethra is short and straight and has a wide caliber and urethritis is therefore almost always a temporary condition However if the periurethral glands are involved they may affect the urethra and bladder permanently Stricture of the urethra is not common in women

The chief symptom of urethritis is burning on urination If there is frequency one suspects some disorder (cystitis) within the urine-containing viscera There may be a constant burning or itching sensation

On examination in the acute stage the urethral meatus is swollen and red and pus can be expressed from its lumen The infection usually involves also the sub mucous tissues and the periurethral ducts of Skene and any glands or congenital diverticula that may be situated along the urethral canal

CHRONIC URETHRITIS Chronic urethritis in the female has no distinguishing features except for the exacerbations of the acute urethritis it originates

On examination the urethra may feel thick and it is sensitive The diagnosis is made by endoscopic examination In addition to the evidence for a chronic inflam mation there are cystic changes in the urethral epithelium

Relaxation of the Vesical Sphincter The vesical sphincter normally is suffi ciently strong to hold urine under excitement coughing sneezing or lifting It is a common experience in parous women that coughing will be accompanied by a loss of a few drops of urine In man and other mammals excessive fear fright or pain will be accompanied by a loss of urine This is called stress incontinence and is a normal act

Stress incontinence of a marked degree may occur in virgins neurotic men and nulliparous women In such instances it is due to an overreaction to which these individuals are subject There is no organic disease of the urinary structures

Childbirth Injuries of the Urethra Childbirth injuries comprise urethrocele relaxation of the vesical sphincter and urethral fistulas

Urethrocele usually accompanies *cystocele* It is a relaxation of the anterior vaginal wall in the region of the urethra The symptoms are identical in both cases Not infrequently a urethrocele remains after the cystocele has been repaired surgi cally unless the urethrocele is discovered and corrected at the same time This may account for the persistence of symptoms after a cystocele has been surgically repaired

Urethral Obstruction Urethral obstruction in women is uncommon It is un usual for a woman to have stricture of the urethra subsequent to gonorrhea or other infections

A urethral obstruction may be partial or complete acute or chronically slow in development depending on its cause

Partial obstruction occurs in middle aged women as a sequel of chronic infec tions resulting from trauma of coitus childbirth or in elderly women from senile changes in the mucous membrane Unusual causes are extensive inflammatory lesions and tumors In middle aged women the obstruction usually is situated in the outer third of the urethra or at the external orifice The orifice is so small that a ureteral catheter is passed with difficulty

The symptoms of partial obstruction are difficult and slow urination burning frequency urgency and pain There is often an associated recurring cystitis As soon as the urethra is completely cured the cystitis usually disappears

In the aged woman who has a small urethral orifice normal functioning may be carried on for a long time The onset of any superimposed inflammatory reaction however soon causes enough edema to transform such a lesion into a complete obstruction

On examination the urethra is more rigid than normal and it feels indurated

usually softer may be attached to the skin by fibrous bands and is often lobulated Occasionally a fibrolipoma becomes pedunculated and may attain large size

Sebaceous cysts are common in the vulvar skin Usually they are round or oval and project above the skin Their characteristic appearance is a grayish color the overlying skin being tense adherent and thin The whole *tumor is freely movable*

Sweat gland tumors (hidradenoma) are uncommon growths that originate in the sweat glands of the labia The tumors ordinarily are not more than ⅖ or ⅘ inch (1 or 2 cm) in diameter They occur in older or at least *middle aged women* The lesions are smooth and covered by normal skin resembling small subcutaneous fibrolipomas or sebaceous cysts Openings in the overlying skin may suggest that the lesion is a sebaceous cyst The histologic appearance may suggest adeno carcinoma

Malignant Tumors *Carcinoma of the vulva* is primarily a disease of women past the menopause although it occurs at times in younger women The average age of the patients is about 60 years These carcinomas are of the squamous cell variety arising from the skin at mucocutaneous borders The lesions appear on the labia on the clitoris or in the vestibule They often are associated with leukoplakia or with carcinoma of the rectum They do not develop rapidly but ulcerate early The lesion becomes a nodular weeping putrid smelling ulcerated mass It is diagnosed by biopsy and histologic examination

Adenocarcinomas of Bartholin s glands are rare They appear as hard nodules beneath the skin in the region of the gland Occasionally adenocarcinomas arise from the sweat glands Secondary adenocarcinomas arising from the uterus or rectum form hard undermining ulcerations in the vulva Diagnosis is by histologic study of biopsied tissues

Melanotic sarcoma and *sarcoma* on the vulva are rare

THE FEMALE URETHRA

The female urethra 1⅗ to 2⅖ inches (4 to 6 cm) long leads from the vestibule to the bladder The external urethral orifice is usually situated about halfway between the clitoris and the anterior margin of the vaginal orifice

It is subjected to and may be injured by the discharges from the uterus and vagina and may be contaminated by feces especially in infants and in adults *who have incom* petent anal sphincters The urethra is easily traumatized in childbirth

Skene s ducts or glands open into the urethra immediately within the external *meatus Other than Skene s glands* Cabot and Shoemaker observed only epithelial crypts and recesses which perhaps were the rudiments of embryonic glandular struc tures but not true urethral glands Urethral diverticula are rare

Hypospadias The female urethral orifice normally is situated well above the vaginal margin and is clearly visible However it may occupy a lower position and open into the vagina This abnormality is due to arrested development of the uro genital sinus and to incomplete separation of the orifices of the urethra and vagina

The extreme malformation is observed when the vagina opens into the *posterior* urethra This is seen only in the presence of gross malformation such as her maphroditism

Hypospadias in women rarely causes any notable symptoms *Occasionally it may* produce external irritation especially on intercourse In some instances this condi tion favors the development and persistence of urethritis and cystitis

Infections of the Urethra *The urethra is often a portal* of infection in girls during childhood Skene s ducts externally and some glands and epithelial crypts in the posterior portion of the urethra may maintain infections and cause recurrences of *cystitis In women* gonococcal infection in diverticula or in Skene s glands may be the focus of reinfection Glands along the posterior part of the urethra and even diverticula are difficult to discover on endoscopic examination

Urethrocele is a herniation through the urethrovaginal septum and not through the urethral meatus

THE INTERNAL FEMALE GENITAL ORGANS

EXAMINATION The examination of the female genital organs is performed with the patient in any one of four positions It is assumed that the patient is fully undressed and that all unnecessary exposure is avoided

1 The routine examination is conducted with the patient lying on her back on an appropriate examining table with her knees flexed and hips fully flexed the thighs drawn up on the abdomen and the coccyx lifted so as to lessen the inclination of the pelvis The heels support the flexed legs on stirrups on the corners of the table or the thighs are supported by leg holders or are held by assistants

2 A position especially valuable for examination of the portio vaginalis the rectum and the bladder is the knee-chest position The elbows and knees of the patient support the body The shoulder girdle is in a low position while the back and buttocks are high The abdominal and pelvis viscera fall toward the diaphragm and the attachments of the pelvic organs are stretched

3 To afford a better view of the cervix the patient lies on her left side with the left arm behind her and allows the right shoulder to sink as far forward or downward as possible the abdomen and thorax resting on the table The knees are flexed and the thighs drawn up (Sims position)

4 The standing posture is especially valuable if the patient has a descensus or other displacement of the pelvic organs For this examination have the patient stand with one foot on a low stool before the examiner In this posture the degree of descensus can be determined and the posterior and lateral walls of the pelvis can be thoroughly palpated

It may occasionally be necessary to anesthetize the patient in order to relax the parts for examination but the greater the experience of the physician the less often will this method be required In young unmarried women a rectal examination will usually suffice

A pelvic examination is never performed without knowing that the bladder is empty For the examination of the female pelvis the index and middle fingers are used The hand may be covered with a rubber glove or the fingers covered with finger cots The covered fingers are lubricated The fingers are introduced into the vagina if the outlet is marital If the outlet is virginal only a rectal examination is made

Bimanual Examination After the preliminary examination of the vagina and cervix the exact position of the *portio vaginalis* or *cervix* is ascertained Normally when the uterus is in the position of anteversion it lies behind the spinal line (connecting the spines of the ischia) the external orifice of the uterus or os being directed backward and the anterior lip being longer than the posterior The cervix should lie approximately in the median line

Bimanually to palpate an anteverted *uterus* the examiner places the fingers of one hand above the right pubic bone and presses gradually into the depth while the fingers in the vagina one finger on the anterior lip of the cervix shove the tissues just in front of the cervix strongly upward As soon as a resistance is felt by the external hand the pressure of both hands may be slightly increased when the body of the uterus can be felt between the two hands Or the fingers in the vagina may press forward from the posterior wall in which event the portio vaginalis along with the body of the uterus is shoved against the external hand Should the uterus lie abnormally high it may be necessary to press with the fingers directly against the tip of the cervix in order satisfactorily to palpate the uterine body

To make sure whether a resistance felt is really due to the body of the uterus a test is made to ascertain whether the portio vaginalis which is certainly a part of

Obstruction by extrinsic tumors is obvious On direct endoscopic examination by one trained sufficiently to perform such an examination the urethra is red and inflamed the normal longitudinal striae are absent and the mucous membrane is injected There may be hypertrophy of the bladder as evidenced by a mucous membrane which is irregular owing to trabeculations of the hypertrophied muscle and inflammation If overdistention and dilatation persist, hydronephrosis and im pairment of renal functional activity ensues

The diagnosis is made by inspection and palpation of the urethra and the passage of urethral sounds and dilators by skilled hands

Incontinence *On occasion in men women and children the bladder gradually* becomes distended with urine because of some neurogenic or organic disturbance in the passage of urine so that it is never completely emptied When the bladder becomes distended to its full capacity and empties only by the pressure of the ab dominal wall the condition is called paradoxical incontinence The overflow voiding *may be practiced by the patient who learns that voiding may be accomplished by* flexion or bending the body forward often while at stool The amount of urine is normal and frequency of urination occurs without intervening incontinence In these patients the full bladder on palpation simulates an abdominal tumor

Enlargements and Tumors of the Urethral Meatus and Urethra *Eversion of the urethral mucosa is common in women because the boundary separating the* mucosa of the urethral canal and the surrounding epithelium of the vestibule is not constant and often this inconstancy is increased by a widely patulous meatus This eversion and redundancy of the urethral mucosa is symptomless and does not require treatment

Prolapse of the urethral mucosa may occur In this condition the urethral mucosa everts itself and projects from the urethral orifice The protruding mass is extremely sensitive and bleeds easily It may be difficult to tell whether the mass is a prolapsed urethral tumor or prolapsed and congested urethral mucosa

Urethrocele or prolapse of the mucous membrane of the urethra has been ob served in girls less than 15 years of age The cause is unknown

Primary tumors of the urethra are rare but secondary tumors are common Carcinomas of the bladder rectum cervix and anterior vaginal wall may extend to involve the urethra

Benign tumors of the urethra are *urethral caruncles* in reality polyps of the urethral mucosa Caruncles are small and are situated near the external meatus They may occur singly or multiply Urethral caruncles are occasionally malignant and therefore biopsy should be performed and they should be treated surgically Carun cles are symptomless until the mass becomes large The symptoms when the caruncle is large are bleeding pain and dysuria On examination there is a dark red mass protruding from or lying in the urethral meatus Biopsy is diagnostic

Urethral polyps are not common and are situated in the middle or the posterior portion of the urethra They have the same features as urethral caruncles The diag nosis is made by the endoscopic examination and when found the polyps are removed

Malignant tumors of the urethra *primary carcinomas* and *sarcomas* like benign urethral tumors are rare They are distinguished from benign caruncles polyps eversion and other lesions by biopsy and microscopic examination

When a mass is found anterior to the vaginal orifice the correct relationship of the urethral canal to the mass is ascertained In a true prolapse the lumen is directly in the center of the mass Occasionally this condition must be differentiated from urethral polyp caruncle condyloma urethrocele and carcinoma In urethral polyps the polyp is in the center of the lumen *and the mucosa is around it In caruncle* the lesion is usually found only on the posterior lip of the meatus and is very sensitive to touch Condyloma has a warty fungoid appearance and is usually multiple

The ovaries are frequently found in an unusual position The ovary is recognized by (1) its position (2) its flattened ellipsoid shape (3) its movability (4) its elasticity and (5) its normal characteristic tenderness

In many adults prolapse of the ovary is found the organ on one or on each side is down in the recto uterine pouch where it may be either loose or fixed Often it is very tender

The vagina and cervix are examined by means of the vaginal speculum For most purposes the bivalve speculum is perfectly satisfactory It is made in several sizes and the examiner selects that size which can be introduced easily In children the most satisfactory speculum is a small cystoscope which is used in the knee-chest position In adults if the hymen is unruptured one may use a very small bivalve speculum

To introduce the bivalve speculum one turns it so that its long axis is vertical to conform to the shape of the vaginal orifice A lubricant facilitates the introduction of the speculum The common practice of using green soap as a lubricant is not advisable on the first examination because soap alters the chemical reaction of the vaginal secretions and interferes with staining and cultural reactions Hence water or a drop of some jelly on the outer surface of the speculum is a better lubricant

THE VAGINA

The vagina is about 3 inches (7 6 cm) long its posterior wall is longer than the anterior being 3½ inches (8 9 cm) long The hollow formed by the anterior wall of the vagina blending with the anterior lip of the cervix is the anterior fornix The depression behind the posterior lip is the posterior fornix behind which is Douglas pouch At the vulvar outlet the lumen of the vagina is anteroposterior in direction it then changes to lateral and at the cervix becomes round Its walls are in contact

Congenital Malformations Absence of vagina absence of uterus double uterus septate uterus bicornate uterus double cervix and double or septate vagina are anomalies due to *inhibition in the development of the mullerian ducts* The ducts are differentiated and attain a certain degree of development then for some reason normal progress is arrested The result is any one of the aforesaid uterine and vaginal malformations depending on the point at which fusion is stopped and the amount of maturity which each mullerian duct acquires independently

When there is *nonunion* of the mullerian ducts the uterus and upper part of the vagina are absent The uterine tubes are separately situated in the lateral parts of the pelvis and each terminates separately and blindly near the midline behind the bladder These mesial ends often form bulbous enlargements 1 cm in diameter which are attached to the round ligaments The defect is usually *limited to the mullerian ducts* so that ovaries external genitalia hymen vaginal orifice and external segment of the vagina are normal

Women who lack the uterus and vagina menstruate into a uterine tube As the result of successive menstruation into the tubes large pelvic masses are formed

Incomplete union of the mullerian ducts permits of a great variety of uterine and vaginal defects depending on the location of the defect and the extent of the inhibition

The uterine defects vary from a very small ridge or depression on the uterine fundus to a complete septum extending through the uterine cavity Bicornate uterus septate uterus and varying grades of duplication may be present One half of the uterus may be normal the other being a rudimentary horn There may be only one cervix there may be two Such defects usually cause no symptoms at all at times pregnancy may develop in a rudimentary horn and may lead to serious consequences

The *vaginal defects* due to incomplete union of the mullerian ducts may result in a vertical septum which completely divides the vagina (septate vagina) These septa do not cause serious inconvenience

Congenital absence or atresia of the vagina may involve the external or the internal part of the vagina

the uterus is connected or not connected with the resistance In addition dependence is placed on the form and consistency of the uterus as felt by the external hand The mobility of the uterus (body and cervix) varies according to size and weight of the uterus the condition of its ligaments the tension of the abdominal walls and the state of the adjacent organs Normally it should be easily possible to press the uterus with its fundus upward as far as the upper margin of the symphysis pubis

The angle between the cervix and the body when the urinary bladder is empty is obtuse in normal anteversion and acute in anteflexion The normal uterus is in a position of antedextroversion the middle of the fundus gravitating slightly to the right

On pressing firmly with the two hands near the lateral margins of the uterus the *broad ligament* (ligamentum latum uteri) on each side may be examined and abnormal conditions of the parametrium will be found if present in the transverse pelvic diameter to the right and to the left of the uterus

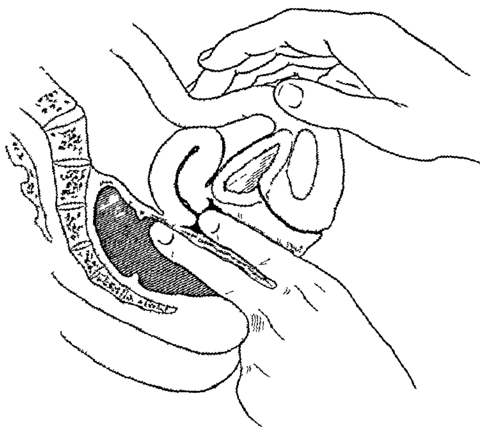


Fig 7 4 Pelvic examination with simultaneous examinations from the rectum and vagina

The *pouch of Douglas* (recto uterine pouch) behind the uterus and the *peritoneal pocket* between the uterus and the bladder (vesico uterine pouch) cannot be demonstrated normally on palpation but if they are filled with blood ascitic fluid inflammatory exudate or tumor the projecting lower part of the pocket can be felt If doubt about the size or position of the uterus exists after the usual pelvic examination the examiner may remove some of this doubt by examining the rectum and vagina simultaneously with one finger in the vagina and the other in the rectum (Fig 7-4)

Dilated and thickened tubes are easily palpable They are distinguishable from other tissues by their elongated shape which they retain even when enlar d

Leukorrhea is a very common complaint of women The discharge may arise in any structure from any or no discoverable cause from Bartholin s glands to the uterine tubes The commonest causes of leukorrhea for which medical aid is sought are normal physiologic variations in the amount of the vaginal secretions and unnecessary douching

A severe vaginal irritation or burn produced by a douche that was too hot is followed by leukorrhea A similar injury may be produced by a douche solution that is too strong or irritating Infections by pyogenic bacteria viruses (herpes simplex) and higher plant (yeast) and higher animal (trichomonas) parasites cause vaginal and cervical irritation and leukorrhea

A yellow fluid discharge is often associated with vaginitis If bubbles are present the infection may be due to *Trichomonas vaginalis* A trichomonad infested vaginal discharge has a foul sour odor

Watery foul smelling vaginal discharge often colored usually contains decomposed blood or organic material This type of discharge was formerly said to be characteristic of cancer of the cervix but it is far too common to have such a diagnostic import

Foreign bodies in the vagina may be the cause of chronic leukorrhea Objects of many different kinds have been recovered from the vagina particularly in the insane and in sexual perverts and thieves Foreign bodies tend to bury themselves in the vaginal wall and may cause considerable pain and discharge They may slough through into the pelvic cavity rectum or bladder They may be so deeply imbedded in the vaginal walls that they can be removed only with the patient under anesthesia Soda pop bottles or similar bottles should be removed from the vagina by warming the bottle to dislodge the mucous membrane which has been sucked up in the neck of the bottle Bottles are inserted immediately after removal of the bottle cap in order to use the contents for a contraceptive douche Bottles become adherent by sucking in the vaginal mucous membrane when the contents of the bottle have been discharged

Cyclic Changes in the Vaginal Epithelium In brief the vaginal mucosa during the lifetime goes through four cycles each of which coincides with the current degree of ovarian activity These four cycles are (1) postfetal or newborn including the first week after birth (2) infancy and childhood from the postfetal period to puberty (3) sexual maturity puberty to menopause and (4) postmenopausal The term senile vaginitis describes the postmenopausal change in the vaginal epithelium

Vaginal Smears Vaginal smears reflect these cyclic changes and perhaps the cyclic changes wrought by the monthly periods with fair accuracy when viewed by a histologist who is an expert in vaginal cellular morphology

Tumors of the Vagina ***Benign Tumors*** *Inclusion cysts* are from some fragment of vaginal mucosa which is buried under the surface by a perineal laceration or operation They are often situated on the posterior vaginal wall Such cysts are difficult to visualize on the anterior wall except when the patient assumes the knee chest position

Inclusion cysts are small cysts less than 1 inch (2 5 cm) in diameter They are easily identified by the bluish color As a rule these cysts cause little if any inconvenience

Endometrial cysts are the result of cystic changes in ectopic endometrium They are situated in the posterior fornix behind the cervix These cysts are a part of the general manifestations of endometriosis elsewhere in the ovaries uterine body or pelvic peritoneum

Gartnerian cysts are formed by cystic dilatation of the remains (Gartner s duct) of the wolffian ducts They are 1 or 2 cm in diameter and are situated on the lateral or anterior walls of the vagina Small cysts rarely cause symptoms when large they

The external type involves only the vaginal orifice hymen or terminal part of the vagina Imperforate hymen is the commonest external vaginal atresia In this type of lesion the menstrual blood accumulates in the vagina (hematocolpos) or in the uterine cavity (hematometra) The collection of menstrual blood forms a soft mass which bulges between the labia In these cases the lesion is due to defective development of the mullerian structures The uterus and uterine tubes are normal

In some instances there may be no external bulging between the labia but a cystic mass is palpable above the imperforate hymen In such a patient the atresia may involve the lower part of the vagina as well as the hymen

When there is an *internal absence or atresia of the vagina* there is usually neither cervix nor uterus but the ovaries are present The external genitalia vaginal orifice and ovaries may appear normal The vaginal orifice is a shallow depression rarely more than 1 inch (about 2 5 cm) deep It may be of normal width or may be very narrow In some instances owing to efforts at coitus or dilatation the vagina may be about 2 inches (5 1 cm) deep

Internal absence or atresia of the vagina is symptomless until the age of menarche or until sexual intercourse is attempted In childhood it may be discovered accidentally by an observant mother or nurse or if accompanied by virilism The persistence of amenorrhea is the first symptom After marriage the complaint is likely to be inability to have coitus (dyspareunia) or sterility The diagnosis is usually evident on examination

Agglutination of the labia minord consists in an adhesion of the proximal edges of the labia The appearance of the anomaly is characteristic There is a bluish discoloration near the center of the membrane formed over the vagina by the labia minora indicating a space below it The membrane is thin and is easily depressed by light touch with a blunt instrument

Hermaphroditism In the human being *true hermaphroditism* is so rare that one should not expect to see it

False hermaphrodites present the genital glands of one sex in an individual whose secondary sexual characters and external genitalia tend to resemble those of the opposite sex The internal genitalia can be those of either sex There may be a doubling or mixing but often some of the parts are atrophic The sex ratio of false hermaphrodites is 3 males to 1 female

All grades of the *male pseudohermaphrodite* exist The general features are clearly or suggestively masculine while the external genitalia are clearly or suggestively feminine In these labial hernias often contain the testicles After puberty the clitoris becomes large it may approach the male penis in dimensions The vagina is usually rudimentary or entirely absent It may open into the urethra The internal genitalia of the male pseudohermaphrodite are often rudimentary

In the female pseudohermaphrodite the gonads are ovaries The external genitalia are of either type although enlargement of the clitoris is not the rule The general external appearance voice bodily contour distribution of hair and temperament are usually female The internal genitalia consist of the ovaries with the accessory male or female organs when present in varying degrees of development

Hermaphrodites may be reared and trained as members of the sex opposite from their gonads During childhood this is serious with the onset of puberty it is often tragic

Leukorrhea The vaginal mucous membrane is protected by variable amounts of secretion to keep its surfaces moist and healthy Excessive but normal increase of vaginal discharges may be present constantly for a time near menstrual periods at the time of ovulation after coitus and occasionally as the result of fatigue excitement or grief Any quantity of objectionable or excessive vaginal discharge is termed leukorrhea Although a white curdlike discharge is normal the normal discharge may be translucent and thin

Childbirth injures the cervical tissues A lacerated cervix may become infected and never heal completely A failure to heal of an obstetric injury an infection or a surgical operation may cause stenosis of the cervix resulting in painful menstruation and sterility

There may be no symptoms of chronic cervicitis Often however there is leukorrhea Rarely does chronic cervicitis give general symptoms

On examination there are areas of redness eversion of the mucosa scars re dundant granulations and dilated glands—all of which are indications of cervical injury and infection

Chronic cervicitis which has followed an unhealed scar may have sharply defined borders or be diffuse to the reflection of the vagina presenting the appear ance of a chronic irritation There may be present yellowish or bluish cystic cervical glands Some cervices are completely distorted by deep scars they may be swollen edematous and enlarged (cervical hypertrophy)

These chronically infected cervices are diagnosed by their appearance on exami nation and by the ease with which they bleed when swabbed with a cotton applicator If bleeding is present it may be well to perform a biopsy of the cervix

Granulomatous Lesions of the Cervix Tuberculosis Incidences of tuber-culosis as reported vary considerably Counsellor and Collins recorded that the patients were 20 to 40 years of age in 73 per cent of their cases and stated that marriage and pregnancy are contributing factors in the disease

Tuberculosis of the cervix is primary if other foci are absent The origin of primary cervical infections is by introduction from the outside This means of infection is exceedingly rare The lesions are secondary to foci in the lungs the upper part of the genital tract and the gastrointestinal tract

Different types of lesions occur The ulcerative type is most commonly seen and appears as a large region of deep ulceration with ragged undermined edges and a red soft easily bleeding granulomatous base Ulcerative and papillary lesions may occur which are not grossly distinguishable from carcinoma

The bleeding may be spotty or profuse Often there is postcoital spotting Leukorrhea is also a prominent symptom the discharge is grayish yellow thick profuse and foul because of the destruction of tissue and the presence of caseous material Loss of weight night sweats anemia and weakness may be present but generally these are due to the presence of tuberculosis elsewhere in the body Pelvic pain is present with secondary infection or with tuberculous extension to the vagina parametrium and upper part of the genital tract

Clinically the diagnosis is seldom made unless *Mycobacterium tuberculosis* is found in the vagina or in the injected guinea pig The *Mycobacterium tuberculosis* cannot be differentiated from the *Smegma bacillus* in stained preparations

Granuloma Inguinale The cervix is one of the primary or secondary sites of granuloma inguinale

The specific etiologic agent is a microorganism which characteristically forms Donovan bodies (Donovania granulomatis) it has not been cultured and is pathogenic only for man The symptoms do not vary from those of carcinoma but the youth of the patient is suggestive of a nonmalignant lesion

On examination the early lesion appears as a shallow ulcerated area with well defined edges and a diffuse red granulomatous base The advanced lesion consists of a large fungiform excrescence of soft red friable tissue replacing part or all of the cervix

The diagnosis depends on a demonstration of the Donovan body in smears scrapings or a biopsy specimen Better results are obtained by the use of biopsy than from smears or scrapings Diagnosis is by no means a simple matter in chronic instances since the characteristic monocytes may be scarce

may interfere with sexual intercourse or with labor The commonest complaint is the foul odor from the interference with normal discharge and drainage of urine

A large vaginal cyst in the anterior fornix may simulate a cystocele A cystocele is emptied by urethral catheterization whereas a cyst is not

Papillomas and *polyps* are small sessile or pedunculated excrescences often multiple and are situated anywhere in the vagina

Condylomas are polypoid growths that are of inflammatory origin Often they are due to syphilis gonorrhea or other infection Pregnancy and obesity favor the development of vaginal polyps and condylomas

Fibromas and *myomas* of the vaginal walls are rare They may occupy any part of the vagina and they occasionally attain large size They are firm movable tumors with a smooth surface

Granulation tissue may occur in vaginal scars Granulation tissue may arise in the margin of the vaginal scar after total hysterectomy These may be carcinomas if the patient has been operated on for carcinoma of the body of the uterus

Malignant Tumors The *carcinomas of the vagina are commonly of* secondary origin They occur in women past the menopause or older

Primary carcinomas may occupy any part of the vagina They are highly malignant ulcerated friable and indurated tumors which are fixed to the subjacent tissues They invade the rectovaginal or vesicovaginal septum or urethra with the formation of fistula urinary incontinence infection pain and incapacitation They metastasize to the inguinal and pelvic lymph nodes The earliest symptom is vaginal bleeding or discharge On examination an indurated fixed ulcerating lesion of the vagina of an elderly woman is usually a carcinoma Biopsy is diagnostic

Secondary carcinomas of the vagina originate from carcinoma of the uterine cervix and uterus Anal and rectal carcinomas often extend to involve the vagina

Sarcomas of the vagina are rare They may be present in infants and in children although no age group is entirely immune Vaginal sarcomas differ from benign polyps in being much harder fixed to the deeper tissues and invasive

THE UTERINE CERVIX

The uterine cervix is the rounded knoblike end of the uterus which opens into the vagina It varies greatly in size and length in nulliparae *It is about 1 inch* (1 to 3 cm) in diameter Occasionally it extends into the vagina as a cone shaped projection 1⅕ to 1⅗ inches (3 to 4 cm) in length The cervical canal opens into the vagina by the external os and into the uterus by the internal os The external os in nulliparae is round but in those who have borne children or in those who have had deep cauterization it is an irregular transverse slit The cervical canal is narrowed at both the internal os and the external os and is larger between hence when instruments are passed into the uterus they traverse with difficulty the external os and the internal os but pass readily between the two and into the uterine cavity beyond

The shape of the cervix and of the external os is so variable after childbirth that *descriptions are impossible Mutilations of the cervix* worse than those of childbirth are rendered to it by surgeons in deeply cauterizing it or attempting to amputate it

Chronic Cervical Irritation (Cervicitis) Many different kinds of organisms cause chronic infection of the cervix Of the bacteria those commonly present in chronic infections are the gonococcus tubercle bacillus and various other pyogenic infectious agents The cervix may be *injured by fungi and Trichomonas vaginalis*

The glands of the cervix may become infected and if so they are often occluded An occluded infection in the cervix frequently becomes enclosed and chronic and persists

grades of malignancy and for this reason length of survival usually but not always coincides with the grade of malignancy revealed by microscopic study

There are no early or first *symptoms* of carcinoma of the cervix There may be irregular bleeding and vaginal discharge In the life of a woman irregular bleeding and vaginal discharges are so common that she is forced to accept these as temporary inconveniences Assuming that all agreed that in an attempt to detect all carcinomas of the cervix early enough for cure biopsy be performed on every woman who has a vaginal discharge and irregular bleeding it would soon be realized that these symptoms are not always present with carcinoma for many women who have carcinoma of the cervix do not have bleeding until the late stages of the disease Some women who have died of carcinoma of the cervix were examined early by those skilled in the art of examination and it was not until symptoms of widespread malignant disease were present that a diagnosis was made This will continue to happen despite all that can be done until more is learned about this disease It is impossible to estimate the stage of invasion by the duration or amount of bleeding Bleeding may occur only after coitus Leukorrhea may come on early in the disease it has no particular diagnostic significance even if the discharge is blood tinged Bloody leukorrhea is more significant of a benign polyp than of carcinoma Pain in the course of cervical malignant disease is an unfavorable symptom since it often indicates an extensive growth

On *examination* the earliest carcinoma of the cervix is superficial limited to the epithelium or may be present in situ and thus it does not invade the subjacent tissue In this stage the lesion cannot be identified by its gross appearance In the early carcinoma of the cervix the malignant change may extend over the cervix or may remain localized near the cervical os For these reasons early carcinomas are discovered only during the microscopic examination of material removed for biopsy

As the growth becomes invasive the appearance of the cervix on examination may reveal that the tumor has grown externally producing an ulcerating mass on the cervix In contrast to this the growth may have extended into the depth of the cervix leaving minimal surface changes However as the tumor develops it extends to the surrounding structures The degree of involvement of the extracervical tissues is important both diagnostically and therapeutically

On palpation there often is extension into the broad ligament and the bladder and posterior extension along the uterosacral ligaments to the sacrum the rectum and the sacral nerve roots The regional lymph nodes are involved in the following order of frequency hypogastric obturator ureteral and external iliac Lymph node involvement is always indicative of extensive malignant disease

The foregoing findings on examination are clearly identified in the *Annual Report on the Results of Radiotherapy in Carcinoma of the Uterine Cervix* (1952) (the so called *League of Nations Classification*) The 12 representatives meeting in session on the occasion of the International and Fourth American Congress of Obstetrics and Gynecology in May 1950 agreed that the staging of carcinoma of the cervix should read as follows (quoted by permission of the Editorial Committee)

Stage O Carcinoma in situ—also known as pre invasive carcinoma intra epithelial carcinoma—and similar conditions

Stage I The carcinoma is strictly confined to the cervix

Stage II The carcinoma extends beyond the cervix but has not reached the pelvic wall The carcinoma involves the vagina but not the lower third

Stage III The carcinoma has reached the pelvic wall (On rectal examination no cancer free space is found between the tumour and the pelvic wall) The carcinoma involves the lower third of the vagina

Stage IV The carcinoma involves the bladder or the rectum or both or has extended beyond the limits previously described

Syphilis A chancre of the cervix in the stage of recognizable development appears as a deep funnel shaped sore with thickened rounded edges which slope down to a smooth base

The macules of secondary syphilis of the uterine cervix appear as grayish white slightly elevated areas Large macules or confluences of smaller ones are termed mucous patches The erosion of the mucous patch is covered with a pinkish or a grayish exudate which may contain spirochetes Secondary syphilitic ulcers are associated with yellow mucopurulent vaginal discharge

The gumma undergoes ulceration and necrosis, or occasionally tissue prolifera tion occurs so that *a fungating soft mass replaces most or all of the cervix*

The diagnosis of primary and secondary lesions depends on the finding of *Treponema pallidum* by darkfield examination since results of serologic tests are not always positive until three weeks after the initial infection The tertiary lesion (gumma) is diagnosed by biopsy and positive serologic tests

Chancroid Chancroid of the cervix is rare Symptoms include soreness in the lower part of the abdomen grayish vaginal discharge and burning and pain in the vagina On examination there may or may not be ulcers of the external genitalia and inguinal lymphadenopathy Pronounced vaginal tenderness is present The cervix appears *to be red and edematous with one or more round firm punched out* ulcerated lesions on the portio vaginalis The ulceration may be covered with an easily removable whitish exudate Diagnosis is established by smear culture skin test and biopsy (see p 410)

Tumors of the Cervix ***Benign Tumors*** *Cervical polyps* usually occur in adult women rarely before the age of 30 years For practical purposes cervical polyps are not malignant but malignant changes occur frequently enough to require histologic examination of every polyp that is removed

Cervical polyps are among the commonest causes of intermenstrual bleeding They may cause leukorrhea and may produce sterility Some cervical polyps if large can be felt on palpation On visualization a cervical polyp appears more reddened and of a deeper color than the cervix It usually projects from the cervical os and can be moved about by means of a cotton swab These polyps often bleed *easily when traumatized They are differentiated from prolapse of the mucous* membrane of the cervix by their mobility and microscopic structure

Since most myomas develop in the body of the uterus *cervical myomas* are rare and they seldom are diagnosed prior to operation

Malignant Tumors *Carcinomas* of the uterine cervix usually occur in women who have borne children and who are between the ages of 40 and 55 years They develop in the regions of chronic irritation epithelial erosion unhealed laceration and in polyps

Macklin studied the morbid heredity in cervical cancer and her conclusions leave little doubt that in certain families but only in certain families does heredity seem to play a part in the incidence of carcinoma Jewish women rarely if ever have malignant cervical polyps and cervical carcinoma

Carcinoma of the external cervix of the uterus arises from the vaginal portion including the transition zone at the external os Carcinoma of the endocervix arises between the internal os and the external os of the cervix

Carcinoma of the cervix has offered difficulties in classification Cervical carcinomas are chiefly of the squamous cell variety A few adenocarcinomas are encountered Either the squamous cell carcinoma or the adenocarcinoma can be classified on the *basis of* anaplasia Broders classification is based on anaplasia In this grouping grade 1 carcinomas the least malignant though definitely malignant show the greatest tendency to approach the normal The cells of grade 2 and grade 3 carcinomas show greater lack of differentiation than those of grade 1 while those of grade 4 are the most ana plastic and therefore are the most highly malignant It is recognized that most cervical carcinomas are *mixed* tumors since they usually contain cells of various types and

The *diagnosis* is established by biopsy If the cervix is chronically irritated or ulcerated in middle aged women and in some young women even if there are no symptoms cancer is suspected

Meigs and Jaffe found in their analysis of the survival rates of patients under going irradiation that most of the deaths occur in the first 2 years and that most patients without obvious disease at the end of 2 years have a good chance for recovery Kimbrough and Tompkins found that 23 3 per cent of the patients survived 5 or more years and that 18 7 per cent of the original group lived more than 10 years after treatment

Bleeding from the cervical stump is a serious symptom for it is most frequently due to carcinoma If cervical carcinoma appears within a year after hysterectomy it was probably present at the time of the operation Biopsy is diagnostic

Many have expressed the belief that carcinoma of the cervix in *pregnant women* progresses more rapidly than in older nonpregnant women It should be realized that a cervical carcinoma tends to cause abortion The presence of carcinoma gen erally is an indication for abortion The religious beliefs of the patient should be respected after the stark facts have been presented to her

THE UTERUS

Attachments of the Uterus In addition to being attached to the vagina the uterus has certain folds or ligaments which pass from it to the surrounding parts Anteriorly the peritoneum is reflected from the uterus at the level of the internal os to the bladder forming the *uterovesical fold* Posteriorly the peritoneum descends from the uterus over the posterior surface of the upper portion of the vagina for about ½ inch (1 3 cm) and thence onto the rectum constituting the *rectovaginal* or *recto uterine fold* The deep pouch so formed is called *Douglas pouch* On each side are three ligaments of which the *broad ligament* is the largest and most important The two broad ligaments and the uterus form a diaphragm which extends from one side of the pelvis directly across to the other thus dividing it into anterior and posterior compartments

At its pelvic attachment the broad ligament widens out having the round ligament as its anterior edge and the *infundibulopelvic* or *suspensory ligament* of the ovary as its posterior edge Immediately posteriorly is the *uterosacral ligament* (recto uterine) it runs from the uterus backward and contains muscular and fibrous tissue the muscular tissue goes to the rectal wall while the fibrous goes to be attached to the second and third sacral vertebrae This ligament on each side forms the outer border of Douglas pouch These ligaments may become sensitive if the so called *uterosacral syndrome* ensues

The round ligament leaves the cornu of the uterus just below and anterior to the uterine tube and passes outward forward and slightly upward to reach the internal inguinal ring and canal through which it passes to end in the subcutaneous tissue and skin of the labium majus pudendi

Congenital Malformations See malformations of the vagina page 449

Chronic Subinvolution Chronic subinvolution of the uterus occurs after childbirth most frequently in women between 35 and 45 years of age However the condition may appear in any woman who has had a baby

The pregnant uterus contains an increased amount of elastic tissue in its walls and in the enlarged blood vessels In the course of normal puerperal involution of the uterus the increased amount of elastic tissue in the myometrium and vessel walls is absorbed and new vessels of smaller caliber are formed When this elastic tissue is not properly absorbed degenerative changes follow The connective tissues undergo liquefaction and hyalinization and become edematous because of failure of re establishment of an efficient circulation The uterus remains enlarged

The common symptoms are profuse menstruation intermenstrual bleeding leukorrhea and pelvic pain On examination the uterine body is diffusely and symmetrically enlarged An exact diagnosis is made only by microscopic study of the excised uterus

This classification was adopted by the three American societies and recommended by the Subcommittee of the World Health Organization on the Registration of Cases of Cancer and their Statistical Presentation

In stage grouping the following general rules should be observed

When allocating a case to a stage nothing but facts revealed by examination should be taken in account

The stage of each case should be decided at examination prior to treatment and this classification should remain The classification may be postponed quite exceptionally and the reasons stated

When it is doubtful to which stage a given case is to be allocated the earlier stage should be chosen

The fact that a single case presents two or more of the conditions which characterize a particular stage does not affect the staging

The Editorial Committee recognizes that the classification of 1950 represents a large step forward but considers that in regard to the uniform presentation of statistics on carcinoma of the cervix there are a few points which need further explanation

1 The type of case to be allocated to Stage O should be strictly defined This must be deferred until further experience on the subject has been accumulated Pending such definitions the collaborators are earnestly requested to allocate to Stage I only such cases in which the presence of an invasive carcinoma is beyond doubt

2 The signs which characterize involvement of the bladder should be defined Cases in which on cystoscopy there is no fistula no ulcerating tumour and no tuberculi visible but which show bullous oedema or ridges and furrows are difficult to classify

The Editorial Committee suggests that

(a) A bullous oedema should only be considered as a sign of involvement of the bladder if the diagnosis is microscopically verified (b) Ridges and furrows should only be interpreted as a sign of involvement of the bladder if they remain during palposcopy This method of examination implies coincident cystoscopy and digital examination of the bladder trigone by rectal palpation Where there is no carcinomatous involvement of the bladder wall the ridges and furrows will disappear when the trigone is lifted up The method has not previously been described It has been used for many years at the Radiumhemmet Stockholm as a routine examination in all cases of cervical carcinoma

3 According to the classification of 1950 a case should be allotted to Stage II if the involvement of the parametrium has not reached the pelvic wall and to Stage III if it has reached the pelvic wall The definitions of 1937 state has not invaded the pelvic wall and has invaded the pelvic wall The Editorial Committee is of the opinion that it should be seriously considered at a future revision of the definitions which of these two versions is more suitable

The study of *vaginal smears* as a means of early diagnosis of cervical or uterine cancer was advocated and practiced by Papanicolaou Cervical cancer is revealed in vaginal smears by the appearance of characteristic cells derived from the superficial layers of the tumor which undergo continual desquamation These cells show great variety of form and size much greater than that seen in sections of the tumor The most characteristic feature of the abnormal cells is the atypical form and structure of their nuclei and vacuolization of the cytoplasm A commonly found characteristic cell type is an extremely elongated one resembling a smooth muscle fiber Erythrocytes are generally found in large numbers Block warned however that a positive diagnosis from a vaginal smear does not indicate immediate radical intervention or irradiation but rather that confirmatory biopsies of the cervix or endometrium should be performed

The commonest cause of spontaneous abortion is the intrinsic factor comprising malformations of the fertilized ovum

There are two groups of *extrinsic* factors which may precipitate spontaneous abortions These are the environmental factors external to the body and the factor of disease within the body For instance it is said that women during the first two or three months of gestation should avoid excesses of drugs alcohol cold heat trauma exertion infections and local stimulation

The diseases within the body which are potent in the production of spontaneous abortion are pyogenic infections gonorrhea and tuberculosis Tumors such as myomas and ovarian tumors may cause abortion Uterine retroflexion laceration and injury to the cervix repeated pregnancies uterine hyperirritability congenital malformations and constitutional diseases all may precipitate spontaneous abortion Diabetes may be associated with abortion If a woman who has an endocrinopathy should conceive the pregnancy is likely to terminate in spontaneous abortion

A general anesthetic drug quinine and castor oil turpentine ergot irradiation and trauma may cause spontaneous abortion All sorts of herbs and folk medicine consisting of pastes and solutions are used for intra uterine injection to produce spontaneous abortion or to bring on the period

In regard to physical exercise it is observed that a healthy ovum in the uterus of a hardworking woman is not dislodged by any sort of ordinary or even laborious task performed by her A diagnostician is loath to attribute abortion to many of the overemphasized inconsequential causes

On the basis of the relative completion of the process abortions are termed threatened inevitable complete incomplete and missed The symptoms diagnostic findings and therapy vary with the type that is present

In a *threatened abortion* the bleeding is slight and the pain if any is only moderate It is a discomfort in the lower part of the abdomen The cervix is not dilated

In *inevitable abortion* the cervix is dilated membranes are often ruptured and bleeding is profuse or has persisted despite rest The uterine contractions return as soon as opiates and rest are discontinued In such cases the process is hastened to completion

In *incomplete abortion* the uterus has been partially emptied spontaneously or intentionally If there are no manifestations of infection it is best to complete the abortion All abortions should be considered to be infected

Habitual abortion has possible endocrine gynecologic and urologic associations However in many instances no disturbances to account for an abortion can be demonstrated in any of these categories

In a *missed abortion* the membranes and the fetus are still in place The fetus is dead There is amenorrhea followed by slight bleeding with some pelvic dis comfort The uterus does not increase in size as the pregnancy lengthens Soon toxic or absorptive phenomena malaise headache nervousness and apprehension supervene The absence of a positive reaction to Friedman and Aschheim Zondek tests or one of these tests for pregnancy would confirm the diagnosis

Therapeutic (Induced) Abortion A therapeutic abortion is performed as a part of the treatment for some disorder that concerns the mother or the fetus The indications for a therapeutic abortion vary so widely that only one rule can be given for the diagnostician to follow This rule is never to advise a therapeutic abortion on data supplied by anyone else Weigh all the findings obtained as the result of personally conducted examinations and from these data make a decision It is well to remember that a therapeutic abortion may carry some risk to the patient depending on her physical condition

Rape resulting in pregnancy is an indication for abortion (see p 479)

Hypertrophy There are women who have diffuse enlargement of the uterus as the result of hypertrophy of the muscle In view of the presence of endometrial hyperplasia in all of these cases it is suggested that the uterine enlargement may be in a measure due to hyperestrogenism

The symptoms are menorrhagia metrorrhagia leukorrhea and pelvic pain On examination the uterus is diffusely enlarged Microscopic study of the uterus establishes the diagnosis

Primary Dysmenorrhea Dysmenorrhea is present in approximately one third of menstruating women It commences with the menarche The distress starts the day prior to or with the menstrual flow and consists of cramps in the lower part of the abdomen which may or may not be associated with backache headache nausea and vomiting It persists through the first or second days of the flow There may be prodromal irritability backache headache and gastrointestinal upsets The dysmenorrhea may partially or wholly cease after the birth of the first child After the age of 30 years in the nulliparous woman the dysmenorrhea may become irregular and of lessened intensity

It is agreed that there is no consistent anatomic lesion in patients who suffer from primary dysmenorrhea It is not due directly or indirectly to a retroverted anteflexed or infantile uterus cystic ovaries presacral neuritis defective uterine musculature or disturbed innervation

The pain experienced is due to uterine contractions This point of view is confirmed by the experimental work of Moir and Bickers working separately and of Randall and Odell Ovulation is a necessary precursor to painful menses The presence of secretory endometrium (suction biopsy) as recorded by Randall and Odell and Sturgis and Albright renders this premise certain Ovulation is prevented by diethylstilbestrol and thus there is no development of secretory endometrium and painless menstruation ensues

The presence of backache nausea vomiting and headache can hardly be explained on an organic basis Premenstrual tension as evidenced by abdominal pain irritability headache backache and nervousness, is on a psychogenic basis These patients should be assured that they are absolutely normal and should receive an explanation in simple terms of the basic physiology of menstrual distress Some relief of pain can be obtained with simple analgesics particularly after the patient realizes the nature of her complaint

Infections of the Uterus Infections of the endometrium are usually a part of a diffuse genital infection Endometritis following childbirth is a serious condition which may leave permanent sequelae Tuberculosis trauma foreign bodies and tumors may cause endometritis

In the acute stages of puerperal sepsis the uterine wall may be severely infected and if uterine drainage is poor a chronic metritis may persist indefinitely

Chronic Metritis The condition occurs in both parous and nulliparous women Often there is an associated inflammatory lesion in the uterine tubes Occasionally chronic endometritis without obvious lesions of the appendages may be encountered Chronic subinvolution and chronic metritis may occur in the same uterus In this event there are the pathologic changes that mark both these conditions The symptoms are menorrhagia metrorrhagia leukorrhea and pelvic pain Examination reveals an enlarged uterus Diagnosis is made by microscopic examination of the uterus

Abortion For diagnostic purposes all abortions are either spontaneous or induced

Spontaneous Abortion The causes of spontaneous abortion (miscarriage) may be divided into two general groups those due to intrinsic and those due to extrinsic factors

the uterus is enlarged and usually smooth in outline If menstrual bleeding has been excessive there is anemia

The diagnosis is made by biopsy of uterine tissue The history and the examination may in some instances be almost conclusive In others not even a suggestion of the true condition will be evident Myoma chronic subinvolution of the uterus and adenocarcinoma of the endometrium are differentiated by endometrial biopsy

External Endometriosis By definition in external endometriosis the endometrium has been transplanted or has originated outside of the uterus (Fig 7 5) An internal endometriosis also may be present and vice versa

The theories of the origin of external endometriosis are (1) embryonic rests (2) metaplasia and (3) transportation of either uterine or tubal epithelium

The theory of embryonic rests of cells derives its plausibility from the fact that epithelium of the uterine tubes and uterus and the germinal epithelium of the ovary all have a common origin in the celomic epithelium In the ultimate distribution and differentiation of the celomic epithelium it is postulated that it may be scattered from the umbilicus to the knees

As the result of irritation inflammation or hormonal disturbances the pelvic peritoneum undergoes metaplasia changing into columnar epithelium and forming glands that resemble endometrium or tubal mucosa in appearance and function

Adult epithelium tubal or uterine may be transported to the ectopic sites by the lymphatics veins and peritoneum or by surgical operation As the lesions progress rupturing of the cysts gives rise to implants throughout the pelvis The fluid from an endometrial cyst is irritating and produces peritoneal reaction with subsequent adhesions cementing the pelvic viscera together The lesions of endometriosis are black or brown puckered or cystic areas The lesions are often situated on the undersurface of the ovary and the adjacent part of the broad ligament and on the posterior surface of the cervix along the attachment of the uterosacral ligaments

Endometrial tissue in or near the umbilicus of the abdominal wall appears as bluish cysts which swell and which may discharge blood with each menstrual period When the lesion is found in an abdominal scar there is a history of some uterine or tubal operation

There have accumulated reports of many instances of endometrial tissue being present in the sigmoid colon Many of the patients who have these sigmoid lesions have suggestive histories and findings of endometriosis only in retrospect after the diagnosis is established by biopsy

The presence of endometrial tissue in the bladder causes frequent and painful micturition

After the menopause whether induced by surgical intervention irradiation or nature endometrial tumors retrogress

Many women who have endometriosis have no symptoms referable to the pelvic organs Generally there have been several years of sterility Symptoms are progressive dysmenorrhea menorrhagia leukorrhea and pelvic discomfort Of these symptoms the acquired progressive dysmenorrhea sterility and menstrual irregularities are significant

On examination if the external lesion like a vaginal or umbilical cyst can be seen it is almost diagnostic in appearance but lesions of this type are rare When cysts are brought into view by means of the vaginal speculum artificial light can be caused to reflect and they shine as do the eyes of an animal at night

On palpation the significant findings are in the adnexa About the cervix and in the rectovaginal septum hard nodular rather fixed lesions are often palpable The ovaries are fixed and enlarged and vary in tenderness The uterus is often retroflexed and in women more than 35 years old is often myomatous

A definite diagnosis is established by biopsy However in a young woman

Endometriosis (Adenomyoma Adenomyosis and Endometrioma) When uterine or tubal mucosa is situated anywhere except in its normal position in the tubes and the uterus the condition is termed endometriosis Endometriosis may be internal (uterine) or external (extra uterine)

The usual and unusual distributions of the lesions are shown in Figure 7 5 The lesions often are multiple occurring in more than one organ The incidence of endometriosis is not known but the disease is common

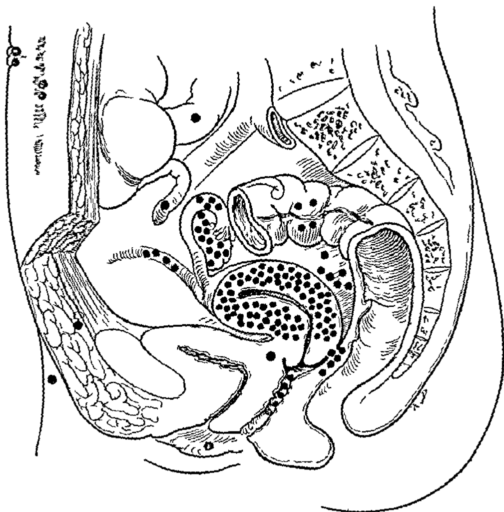


Fig 7 5 Endometriosis The commonest sites of occurrence are indicated

Internal Endometriosis When endometrium is found disseminated throughout the uterine wall the condition is termed internal endometriosis In internal endometriosis the epithelial tissue is continuous with the endometrium which lines the uterine cavity

The ectopic endometrium is scattered throughout a part or all of the uterine wall When the epithelial elements are collected into nodules the condition is termed endometrioma When these epithelial elements are in muscle tissue they are termed adenomyoma

The first symptom of uterine endometriosis is a period of infertility followed by symptoms of pressure of the enlarged uterus and menstrual irregularities The menstrual irregularities comprise a progressive dysmenorrhea and menorrhagia The menstrual periods are often accompanied by intense pain On pelvic examination

advanced pregnancy is detected by the presence of fetal movements and fetal heart sounds and by roentgenologic evidence

Endometrial cysts chocolate cysts and tubo ovarian masses simulate uterine myoma and are often associated with fibroid tumors of the uterus Small asymptomatic myomas do not require treatment Large tumors should be removed

Malignant Tumors *Carcinoma of the endometrium* occurs more frequently in women after the menopause than in younger women The first symptom is frequently postmenopausal bleeding If the tumor occurs prior to the menopause intermenstrual bleeding and profuse menstruation occur In the beginning the bleeding is slight and transitory and may never be more than an intermittent brownish or bloody tinge in leukorrhea which has been present for a long time Leukorrhea is not significant As the growth advances however the discharge may become more profuse and foul On examination the uterus is somewhat enlarged and soft The diagnosis is made by microscopic examination of the endometrium

Papanicolaou became aware that carcinoma of the fundus of the uterus as well as carcinoma of the cervix (see p 456) are to some extent exfoliative lesions in the sense that cells at the free surface of the growth tend to become dislodged and subsequently to find their way into the vagina He developed a method for collecting the cellular debris from the uterus as well as the cervix which is smeared on glass slides and stained in a particular way so that the various components may be studied The method is simple and may be applied to large numbers of women Papanicolaou states that cells pathognomonic of cervical and fundal carcinoma can be definitely recognized The interpretation of smears calls for an intimate knowledge of the cytologic characteristics of the vaginal fluid

Malignant cells will not always be present in every smear from cases of cancer even though the tumor may be very large and clinically obvious The method seems to be used as an adjunct rather than a substitute for biopsy and histologic examination

Primary sarcoma of the uterus is very rare When present it occurs in the walls of the uterus

Secondary sarcoma in a myoma is rare The frequency of sarcomatous changes in myomas is not known

When sarcoma is present in a myoma there is rapid growth and often pain These rapidly growing myomas may occur after the menopause when myomas should decrease in size The examination reveals a tumor that feels like a myoma

The diagnosis is not made until the tumor is examined histologically

Tumors of the Chorion **Benign Tumors** A *hydatid mole* (hydatidiform mole vesicular mole) is a rare lesion When it occurs it follows pregnancy by several months to 2 or 3 years

The hydatid mole may invade the entire thickness of the uterus and perforate the peritoneum This may produce peritonitis and intraperitoneal hemorrhage

Embolic transplantation of fragments of hydatid mole tissue has been observed in the vaginal walls the parametrium and the lungs Transplantation of this tissue does not indicate metastasis as is observed in malignant disease A certain percentage of benign hydatid moles however become malignant chorionepitheliomas

Intermittent uterine bleeding is a symptom There are periods of active bleeding between which a serosanguineous discharge appears Soon there are pain in the lower part of the abdomen weakness and often anemia Abortion is frequent The abortion is accompanied by hemorrhage which is usually profuse prolonged and sometimes fatal because uterine muscle may be invaded and unable to contract vigorously

On examination the uterus seems to be pregnant although there are no evidence of the fetus no fetal heart beats no fetal movements and no fetal shadow on roentgenographic examination Reactions to the Friedman and the Aschheim

who gives no history of a pelvic infection and who has hard nodular lesions about the cervix and retrovaginal tissues with fixation of the ovaries in association with sterility and progressive dysmenorrhea the diagnosis is reasonably certain to be endometriosis

In the differential diagnosis consideration is given to (1) chronic salpingitis (2) uterine myomas (3) the presence of ureteral renal and ureteral stones and (4) lesions of the pelvic colon

Endometriosis of the rectovaginal septum is distinguished from carcinoma by the presence of endometriosis elsewhere the absence of *metastasis and presence* of endometrial cysts behind the cervix

Uterine Neoplasms ***Benign Tumors*** The myomas the fibromas or the myofibromas comprise the major part of the important benign tumors of the uterus

A *uterine myoma* is a benign *tumor composed of smooth muscle and connective* tissue This type of tumor is known also as fibromyoma myofibroma leiomyoma and fibroma The etiology is unknown

Myomas are common in women of all races They are most frequently found in women from 40 to 45 years of age although they may occur in young women the age limits are usually given as from 30 to 55 years Uterine myomas have a wide range of size situation and shape

Myomas are often referred to according to their situation for instance as intramural subperitoneal or submucous The large myomas show hyaline and cystic changes Suppuration and formation of abscess are rare Incomplete or complete calcification may occur Intramural myomas may cause intramenstrual bleeding

Generally uterine myomas are symptomless Symptoms do occur however, and *when present are due to changes within the myomas to degenerations to* unfavor able anatomic relationships and to submucous development The common symptoms are pain tumor menorrhagia and sterility The patient may first palpate the mass which may reach the umbilicus before there are symptoms More frequently how ever a large myoma is sensitive to pressure There is pain which is due to a com plicating adnexal or pelvic disease The pain may be a sensation of pressure or dysmenorrhea dysuria painful defecation or a sacral backache Occasionally it is referred down the thighs or up into the ureteral or the renal regions

Dysmenorrhea is often associated with profuse menstrual bleeding

On routine pelvic examination of women who have attained the late thirties or more a few of them by no means all will have one or more myomas The myomas are an insensitive uterine enlargement which is nodular hard and movable The masses are usually round and multiple and vary in size up to those which occupy *the entire abdomen A large myoma which* has undergone degeneration may be soft and cystic and cannot be distinguished on palpation from a normally pregnant uterus or an ovarian cyst Small myomas which are clustered about the uterus cannot be differentiated by physical means from an endometriosis

Anemia may result from the menorrhagia and may be serious Occasionally anemia is encountered in women who have myomas but who have not had bleeding These anemias are the same as those present in many women who menstruate These anemias can be corrected by the administration of *iron or the patients* may spontaneously recover after hysterectomy

In diagnosis a tumor situated in or adherent to the uterus without symptoms is usually a myoma and such a tumor associated with the forementioned symptoms is nearly always a myoma

A soft myoma causing symmetric enlargement of the uterus is indistinguishable on palpation from a normal pregnant uterus The Aschheim Zondek Friedman and frog tests are the most certain methods of detecting an early pregnancy A *more*

LOCALIZATION The vaginal epithelium in children in women during pregnancy and in senile women is thin its secretions are but slightly acid and it offers slight resistance to the gonococci

The vaginal epithelium of the adult nonpregnant woman is thicker produces a stronger acid secretion and is resistant to gonococci In young women gonorrheal vaginitis is usually slight and transient and there is no localization in the vagina and cervix as is observed in girls and in aged or pregnant women Gonorrheal endometritis is generally transitory

In the periurethral ducts of Skene and the vulvovaginal and cervical glands the gonococcus grows and produces its full effects There are swelling and inflammation which occlude the openings to the ducts and result in acute chronic deep and permanent infections and formation of abscesses and cicatrices If the tubal openings are closed by the inflammation pyosalpinx and hydrosalpinx are often present Sterility recurring exacerbations of the infection pelvic peritonitis and pelvic abscess are serious complications resulting from gonorrheal salpingitis If the tubes remain open as they usually do no serious sequelae are observed As a rule a localized pelvic peritonitis accompanies salpingitis

Ophthalmia when it ocurs is usually a direct contamination transmitted from the genital tract to the eyes by the hands

Among metastatic infections (lymphogenous or hematogenous) are adenitis septicemia endocarditis hepatitis arthritis periostitis osteomyelitis tenosynovitis and dermatitis All of these are described under their respective headings elsewhere in this text

The chronic foci of infections in the external genitalia are the cervical glands and epithelium Bartholin s glands and Skene s glands In children and in the aged or the pregnant woman the vagina is the focus of chronic infection

The infection becomes chronic by closing or constricting the ducts of the glands This interferes with the escape of infectious material for long periods of time The material which collects thus may form an abscess In time if the abscess resolves the fluid in the occluded gland may be clear or translucent the gland being changed into a cyst This cyst in turn may become an acute pyogenic abscess or may remain a cyst The size of the abscess or cyst varies occasionally it attains several centimeters in diameter The orifices of Bartholin s glands are occasionally just visible as red spots (gonococcal maculae) In ancient infections Bartholin s glands remain indurated

SYMPTOMS The symptoms of gonorrhea are classed as acute and chronic Many women during acute gonorrhea notice a slight burning on urination perhaps an increase of a vaginal discharge and discomfort These symptoms are experienced frequently by most women however and are usually inconsequential Other women have had no unusual symptoms and are unaware that anything is wrong until salpingitis develops A few women have acute symptoms comprising definite burning on urination frequency vaginal pain itching and leukorrhea

Chronic gonorrhea of the external genitalia of women as a rule does not cause symptoms In a few women however the symptoms of chronic gonorrhea of the internal genitalia never seem to subside There are leukorrhea and urinary symptoms and a continuance of the active and infectious disease In a few cases cysts of Bartholin s glands may cause local inconvenience recurring abscesses of Bartholin s glands cause local pain swelling and redness recurrent burning on urination and cystitis Sterility may be the complaint caused by a gonorrheal cervicitis However sterility does not often occur for many women who have chronic gonorrhea bear children normally

After long periods without symptoms there may be exacerbation of the infection abscess of Bartholin s glands cystitis salpingitis or arthritis

EXAMINATION In the acute stage there are general swelling and redness of the genital mucosa accompanied with a purulent discharge from the urethra and cervix Results of examination of those who have chronic gonorrhea may be negative or there may be pus expressed from Skene s glands

Zondek tests are often *stronger than normal in a live hydatid* Diagnosis is established by microscopic examination of the products of uterine curettage

Malignant Tumors Chorionepithelioma a rare complication of a rare disease the hydatid mole may follow abortion full term pregnancy or ectopic pregnancy or may occur as a tumor of the testicle In men it exhibits the same degree of malignancy and high urinary titer of hormones (Aschheim Zondek Friedman and frog tests) that are observed in women Metastasis in chorionepithelioma is wide spread and early The common sites of metastatic lesions are the lungs, vagina and brain

The appearance of a vaginal lesion may be the first indication of a chorionepi thelioma After an abortion full term pregnancy or ectopic gestation repeated or continued bleeding usually is due to *retained placental or decidual tissue* and curettage is performed If bleeding recurs after curettage and is accompanied by a rising titer of chorionic hormones in the urine the diagnosis of chorionepithelioma can be *made from these symptoms The diagnosis is established by microscopic* examination of a metastatic nodule or the primary tumor

THE UTERINE (FALLOPIAN) TUBES

A normal uterine tube is about 4½ inches (11 4 cm) long and runs in the broad ligament along its top or free edge from the uterus to the ovary

CONGENITAL MALFORMATIONS The uterine tubes develop from the mullerian ducts along with the uterus and the upper part of the vagina In a grave defect such as complete nonunion or marked inhibition in the development of the mullerian ducts the uterine tubes are likely to be rudimentary In the absence of the uterus or in the presence of completely double uterus or rudimentary uterus the uterine tubes are hindered from their normal development

Double Fimbriae This condition is not rare and is inconsequential causing no symptoms

Congenital Occlusion Congenital occlusion of the uterine tubes is accompanied by *retarded uterine development (hypoplasia)*

Absence of Uterine Tube This malformation is rare It is usually due to aplasia of an entire mullerian duct and is evident in the resulting type of uterine deformity *uterus unicornis In this condition the entire uterus* cervix and upper part of the vagina are formed from one mullerian duct

Absence of one mullerian duct with consequent absence of one tube may be accompanied by absence of the homolateral ovary and kidney

Rudimentary Uterine Tube This is also rare except in the presence of rudimentary development of the whole mullerian duct

Infections Infections termed salpingitis are the most important lesions of the uterine tubes They are significant not only because of their effect on the patient s health but also because they frequently produce permanent tubal occlusion and sterility

The inflammatory diseases of the uterine tubes conform to certain definite types of which the commonest are those due to (1) gonorrhea (2) pelvic abscesses (3) tuberculosis (4) puerperal infections and (5) appendicitis Other less common varieties are associated with systemic infection Not all tubal infections are discussed here

1 ***Gonorrhea*** Gonorrhea is primarily an infection of the mucous membrane of the urogenital tract of men women and girl children In women of the child bearing age the characteristic type of infection occurs in the *uterine tube* It is for this *reason that* gonorrhea in women is discussed with diseases of the uterine tubes the epithelium of the uterine tubes offers little resistance to the gonococci

Since the advent of chemotherapy and antibiotic therapy the following sequence of events *occurs only* when the infection exists without the benefit of the use of these drugs

LOCALIZATION The vaginal epithelium in children in women during pregnancy in senile women is thin its secretions are but slightly acid and it offers slight stance to the gonococci

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DIAGNOSIS The presence of gonococci in these secretions demonstrated by smears by cultures or both establishes the diagnosis However in many cases the diagnosis is impossible The gonococcus is so deeply buried in glands and epithelium that it is inaccessible Positive results on smears or cultures may be obtained during or near the menstrual periods

a **Gonorrhea of the Uterine Tubes** (*Salpingitis*) Salpingitis occurring at the time of a menstrual period or after childbirth or miscarriage may be the first manifestation of gonorrhea of the uterine tubes Many times salpingitis has followed intracervical or intra uterine instrumentation

SYMPTOMS During a menstrual period complicated by salpingitis the dysmenorrhea if present is increased the period is prolonged and the flow more profuse After the period the pain continues and increases Micturition is painful Leukorrhea fever backache painful defecation anorexia nausea and often vomiting are not unusual

In some instances the infection is severe and if proper antibiotic therapy is not given recovery may be delayed by formation of abscess and exacerbations of symptoms during successive menstrual periods Acute abscesses occur in the uterine tubes or the ovaries (pyosalpinx ovarian or corpus luteum abscess) in the pelvis (pelvic abscess) Bartholin s glands urethral glands or perirectal tissues However if antibiotics are administered these complications can be minimized or prevented

One attack of acute salpingitis even without treatment does not necessarily indicate that more attacks will follow The patient may recover completely or partially without treatment In the severe infections complete recovery may be assumed when there is restitution of normal childbearing and menstrual functions The internist is in no hurry to advise surgical operation on a young woman who has acute or subacute salpingitis simply because recovery is slow

EXAMINATION The abdomen may be moderately distended and there is bilateral tenderness in the suprapubic region On occasion the tenderness is limited to the right lower quadrant requiring a differentiation from acute appendicitis

Muscle guarding is infrequent in salpingitis Often a mass is demonstrable early in the course of the disease

In many patients who have acute salpingitis a pelvic examination reveals evidence of gonorrhea of the external urogenital tract On bimanual palpation the characteristic findings are bilateral tenderness and increase in the local temperature of the parts The pelvis is generally and bilaterally sensitive and movements of the cervix and uterus cause pain Often no masses are palpable at this stage There may be only a general sense of resistance and tenderness through the whole pelvis

There may be found thickening of the ovarian capsule and adhesions between the ovary uterine tube and surrounding organs Perioophoritis ovarian abscess tubo ovarian cysts pyosalpinx and tubo ovarian abscesses are common findings

Thickening of the ovarian capsule and its adhesions causes impediment of ovulation by ovarian adhesions and may result in graafian follicular cysts If so when the graafian follicle ruptures virulent gonococci may penetrate the corpus luteum and cause a corpus luteum abscess

The adnexa may be adherent to the posterior surface of the uterus and intestines and omentum may become adherent to the fundus but infection is superficial Intramural abscesses have occurred The endometrium is almost immune to the gonococcus except when traumatized

Laboratory tests may reveal leukocytosis with an increase in the ratio of the polymorphonuclear cells The sedimentation rate is increased

DIAGNOSIS Positive smears and cultures for gonococcus are the only means of establishing a positive diagnosis The history and the findings on examination are usually sufficient for a clinical diagnosis

Acute salpingitis may closely simulate acute appendicitis tubal pregnancy and ovarian cyst with twisted pedicle and it may resemble many other pelvic conditions also Chronic salpingitis may simulate almost any pelvic disease With its many pelvic lesions chronic salpingitis often presents atypical features which may be confusing or impossible of exact diagnosis until biopsy has been made or positive culture for the gonococcus is obtained

b ***Gonorrhea in Pregnant Women*** During pregnancy gonorrhea seems to be able to produce a more serious vaginitis than when pregnancy does not exist After the uterus has been emptied during the puerperium or after the first menstrual period after the puerperium severe salpingitis often occurs

c ***Gonococcal Vaginitis in Children*** Gonococcal vaginitis is common where ever persons who are infected with the gonococcus come into contact with girls Gonorrheal infection is conveyed by infected hands or by infected toilet seats or other bathroom facilities Gonorrheal vaginitis may attain epidemic proportions in orphanages of girl children

The infection causes a deep inflammation which extends through the epithelial layer into the fibromuscular tunic of the vagina There is vulvovaginitis and the labia minora and the inner surface of the labia majora are swollen and red The chief symptom is a purulent vaginal discharge There are discomfort itching and burning on urination

The course of the disease before the era of antibiotics and chemotherapy was most intractable and persistent Many patients were not cured until they reached the menarche when the infection was eliminated physiologically by the natural development of a new type of vaginal epithelium

The diagnosis is established by identification of the gonococcus by smear or culture The presence of a purulent discharge in a girl is suggestive of gonorrhea

d ***Gonococcal Peritonitis*** An acute gonococcal salpingitis is constantly accompanied by an acute pelvic peritonitis which is limited to the pelvis and disappears spontaneously in a week or 10 days It produces an acute general peritonitis in children and young girls which is often the first indication of gonorrhea Occasionally an acute peritonitis may arise from a chronic salpingitis General peritonitis secondary to chronic tubal infection may develop as the result of rupture and escape of infectious material from a tubo ovarian abscess or pelvic abscess

2 ***Pelvic Abscess*** Pelvic abscesses are multilocular and those intrinsic to internal genitalia of the female communicate with tubo ovarian abscesses or a pyosalpinx Often they are extrinsic to the pelvic organs and are due to the col lection of infected fluid from the rupture or chronic perforation of some intra abdominal viscus

Pelvic abscesses are enumerated according to the close association and frequency of infection of the structures in and around the pelvis (1) postappendical (2) salpingitis (3) postoperative collection of serum or pus (4) pelvic abscess from perforation of sigmoid or cecum (5) postoperative hematoma or hematocele (6) infected hematocele from rupture of ovarian cyst (7) osteomyelitis (8) diver ticulitis of the pelvic colon (9) chronic ulcerative colitis or ileitis and (10) puerperal infections

Pelvic abscesses may rupture spontaneously into the bladder vagina rectum or sigmoid They may rupture through the abdominal wall particularly those which arise from chronic ileitis Abscesses from chronic ileitis and from puerperal infec tion may reach the extraperitoneal fascial planes and burrow around the bladder between the layers of the broad ligaments down the fascial planes of the thigh or pass upward into the perirenal fossae A pelvic abscess may follow the urachus to the umbilicus and may rupture and drain at that point

Fistulas follow the rupture of the abscess into the vagina bladder or rectum

DIAGNOSIS The presence of gonococci in these secretions demonstrated by smears by cultures or both establishes the diagnosis However in many cases the diagnosis is impossible The gonococcus is so deeply buried in glands and epithelium that it is inaccessible Positive results on smears or cultures may be obtained during or near the menstrual periods

a ***Gonorrhea of the Uterine Tubes*** (*Salpingitis*) Salpingitis occurring at the time of a menstrual period or after childbirth or miscarriage may be the first manifestation of gonorrhea of the uterine tubes Many times salpingitis has followed intracervical or intra uterine instrumentation

SYMPTOMS During a menstrual period complicated by salpingitis the dys menorrhea if present is increased the period is prolonged and the flow more profuse After the period the pain continues and increases Micturition is painful Leukorrhea fever backache painful defecation anorexia nausea and often vomit ing are not unusual

In some instances the infection is severe and if proper antibiotic therapy is not given recovery may be delayed by formation of abscess and exacerbations of symptoms during successive menstrual periods Acute abscesses occur in the uterine tubes or the ovaries (pyosalpinx ovarian or corpus luteum abscess) in the pelvis (pelvic abscess) Bartholin s glands urethral glands or perirectal tissues However if antibiotics are administered these complications can be minimized or prevented

One attack of acute salpingitis even without treatment does not necessarily indicate that more attacks will follow The patient may recover completely or parti ally without treatment In the evere infections complete recovery may be assumed *when there is restitution of normal childbearing and menstrual functions* The internist is in no hurry to advise surgical operation on a young woman who has acute or subacute salpingitis simply because recovery is slow

EXAMINATION The abdomen may be moderately distended and there is bi lateral tenderness in the suprapubic region On occasion the tenderness is limited to the right lower quadrant requiring a differentiation from acute appendicitis

Muscle guarding is infrequent in salpingitis Often a mass is demonstrable early in the course of the disease

In many patients who have acute salpingitis a pelvic examination reveals evidence of gonorrhea of the external urogenital tract On bimanual palpation the characteristic findings are bilateral tenderness and increase in the local temperature of the parts The pelvis is generally and bilaterally sensitive and movements of the cervix and uterus cause pain Often no masses are palpable at this stage There may be only a general sense of resistance and tenderness through the whole pelvis

There may be found thickening of the ovarian capsule and adhesions between the ovary uterine tube and surrounding organs Perioophoritis ovarian abscess tubo ovarian cysts pyosalpinx and tubo ovarian abscesses *are common findings*

Thickening of the ovarian capsule and its adhesions causes impediment of ovulation by ovarian adhesions and may result in graafian follicular cysts If so when the graafian follicle ruptures virulent gonococci may penetrate the corpus luteum and cause a corpus luteum abscess

The adnexa may be adherent to the posterior surface of the uterus and intestines and omentum may become adherent to the fundus but infection is superficial Intra mural abscesses have occurred The endometrium is almost immune to the gono coccus except when traumatized

Laboratory tests may reveal leukocytosis with an increase in the ratio of the polymorphonuclear cells The sedimentation rate is increased

DIAGNOSIS Positive smears and cultures for gonococcus are *the only means* of establishing a positive diagnosis The history and the findings on examination are usually sufficient for a clinical diagnosis

entiation of genital tuberculosis from chronic salpingitis is impossible until the tissue is studied histologically

Of the various methods used to establish the definite nature of the adnexitis curettage is the one most widely used although exploratory laparotomy salpingo graphy and culdoscopy are also occasionally employed Curettage is occasionally followed by exacerbation of the tuberculous process Inoculation of menstrual blood into guinea pigs can be valuable in establishing the tuberculous nature of salpingitis

Extra uterine Pregnancy The situation of extra uterine pregnancy most commonly is the *uterine tube* rarely the ovary or the abdominal cavity Ectopic gestation often occurs with the first pregnancy if with the second pregnancy or successive pregnancies a period of sterility may precede the ectopic gestation

When the fertilized ovum is prevented from passing through the uterine tube extra uterine pregnancy results Salpingitis is the commonest cause of tubal pregnancy but other causes may be present such as peritubal adhesions or a congenital narrowing Other causes of tubal obstruction are diverticula rudimentary horn of double uterus and pelvic tumors

The sequence of the symptoms may be a preliminary amenorrhea or delayed menstruation followed by irregular bleeding and sudden or repeated pain There may have been slight anorexia and nausea swelling of the breasts and mild pelvic discomfort Severe pelvic pain feeling of weakness or faintness fainting nausea vomiting and vaginal bleeding shock and collapse may develop rapidly In about 1 in every 4 sudden collapse does not occur Patients who do not have sudden collapse may feel only a sense of profound weakness faintness dizziness and incapacitation In some instances the patient may suspect that she is having a mis-carriage

As the amount of blood in the pelvis increases and adhesions form the pelvic discomfort is increased by an exertion defecation and micturition

On examination he pelvis is so sensitive and the patient in such severe shock that the ruptured tube is not palpable blood crepitus is usually absent

In the less acute tubal ruptures examination reveals a normal abdomen On pelvic examination the cervix is often softened and the uterus is soft and somewhat enlarged The characteristic feature is a soft sensitive cylindrical mass in an adnexal region or in the recto uterine pouch On rare occasions if manipulation is not considered too dangerous the ovary on the same side may be isolated and if so the tentative or presumptive diagnosis of unruptured tubal pregnancy may reasonably be made

In tubal pregnancy as long as the fetus or the trophoblast survives reaction to the Aschheim Zondek or a similar test is positive After the fetal elements die the reaction becomes negative unless there is a proliferative independent growth of the trophoblast such as occurs in the presence of hydatidiform mole or unless there is also an intra uterine pregnancy

A specimen of urine is obtained and examined for pregnancy hormones If results are positive examination under anesthesia is indicated and if the findings are confirmed laparotomy is performed

In the acute types of tubal rupture the *diagnosis* is established by the history and the acuteness of the onset and collapse In the less acute type of tubal pregnancy without acute symptoms but with repeated attacks of moderate discomfort the diagnosis is much more difficult The condition may simulate threatened abortion appendicitis salpingitis ovarian cyst with torsion of the pedicle ruptured corpus luteum cyst and a variety of pelvic and abdominal conditions

Tumors *Papilloma* of the uterine tube causes no symptoms It is the only common benign tumor of the tube Cysts in the broad ligament remnants of the paroophoron may be difficult to distinguish from hydrosalpinx

The fistulas from abscesses of the pelvic organs often heal spontaneously Fistulas from ruptured diverticula of the colon and those from chronic ileitis never heal spontaneously

In the acute pelvic abscess the symptoms are those of acute salpingitis The fever is high the pain intense and the pulse rapid In chronic pelvic abscess there may be no acute symptoms The hard mass is found on pelvic examination made because of vague pelvic complaints

If the patient who has a pelvic abscess suddenly begins to have acute abdominal pain with nausea and vomiting there may have been a rupture of the abscess into the abdominal cavity with subsequent peritonitis The abscess should be evacuated surgically

Abdominal palpation discloses tenderness and perhaps a mass in the lower part of the abdomen Pelvic examination reveals a mass and there may be fluctuation in the recto uterine pouch The indurated adnexa may or may not be identifiable As a rule in the presence of a pelvic abscess intrapelvic structures cannot be identified

Chronic pelvic abscesses are hard and fixed masses moderately sensitive and situated behind the broad ligaments

On rectal palpation the finger is insinuated higher in the pelvis than it can be through the vagina The finger can often determine the extent of the abscess its relation to the pelvic walls the cervix and the rectum The presence of pus is indicated by anemia long standing illness with the history of fever and infection and cachexia

3 ***Genital Tuberculosis in Women*** The pelvic organs are usually infected with tuberculosis through the blood from distant foci such as lungs the mediastinal or peribronchial lymph nodes the intestinal tract peritoneum or bones The infection occurs usually between the ages of 20 and 40 years In adolescent girls the disease often takes the form of a diffuse miliary tuberculous peritonitis with ascites Aged women occasionally succumb to the disease Genital tuberculosis is rarely contracted by coitus The uterine tubes are nearly always involved whereas the ovaries are involved in only about a third of the cases and the uterus (endometrium) in from one half to three fourths of the cases Clinically tuberculous salpingitis does not differ from other forms of adnexitis Almost always tuberculosis affects both uterine tubes Except for isolated instances of vulval cervical and vaginal tuberculosis genital tuberculosis in women means involvement of the tubes

In 1 of every 4 women who have genital tuberculosis there is active pulmonary tuberculosis Active pulmonary lesions are therefore important symptomatically and diagnostically

A long history of chronic constitutional and gastrointestinal symptoms accompanied by menorrhagia dysmenorrhea amenorrhea intermenstrual spotting leukorrhea sterility and recurring pelvic and abdominal pain is suggestive of genital tuberculosis If these symptoms are associated with pulmonary manifestations of tuberculosis they are more significant

If on examination of a virgin there are found pelvic lesions indicative of chronic salpingitis adnexal mass induration fixation of the adnexa and unexplained tenderness a presumptive diagnosis of genital tuberculosis is made

In a married woman absence of signs of gonorrhea and no reasons to suspect gonorrhea would support the diagnosis of tuberculosis if there was present evidence of a chronic salpingitis

In the presence of an active pulmonary tuberculosis a pelvic inflammatory disease may be of tuberculous origin If the cervix is involved biopsy may reveal the presence of tuberculosis In the absence of active pulmonary lesions the differ

Small follicular cysts do not cause symptoms A large cyst or many small cysts in the ovaries may produce pelvic pain menorrhagia and at times amenorrhea On pelvic examination the ovaries are bilaterally enlarged tender and cystic The diag nosis thus is strongly suspected A definite diagnosis may require time and finally histologic studies

Corpus Luteum Cysts These cysts may develop rapidly Once formed they may remain for a long time

Pain which is not characteristic and menstrual disturbances are present when there are corpus luteum cysts The menstrual disturbances are the usual ones menorrhagia metrorrhagia and amenorrhea At times these cysts rupture and permit of severe intra abdominal bleeding and the symptoms then produced simulate those of abortion or tubal pregnancy Hematomas contained within the cysts in the pres ence of salpingitis may become infected and form abscess (corpus luteum abscess)

Lutein cysts are usually bilateral and multiple and constitute a mass that may be up to 4 inches (10 2 cm) in diameter Ovarian cyst is about as accurate a diag nosis as can be made

Cystomas The ovarian cystomas comprise the simple cystomas the serous cystadenomas and the pseudomucinous cysts These cysts are histologically benign tumors

The simple serous cystomas may be unilateral or bilateral They are unilocular cysts with thin walls and contain clear fluid The serous cystadenomas are filled with a watery fluid The pseudomucinous cystadenomas are multilocular The con tents of these cysts are a thick glairy gelatinous material that contains pseudomucin

The *serous cystadenomas* may occur unilaterally or bilaterally They attain great size In the presence of pseudomucinous cyst fatal results may follow dissemination of pseudomucinous material in the peritoneal cavity during a surgical operation This condition is known as *pseudomyxoma peritonaei* There is a rapidly accumulat ing ascites

On examination the cystomas are found as bilateral lobulated cystic tumors On palpation these cysts are not uniform some portions of the surface are very soft and collapsible owing to low tension of the enclosed fluid others are hard and resistant giving the impression of almost complete solidity owing to the glandular growth in the wall of the cyst

These different cysts cannot be said to be benign or malignant until seen and examined microscopically

Tumors Arising from Embryonic Rests of Cells The tumors of the ovaries which arise from embryonic rests are the granulosa cell tumors arrhenoblastoma adrenal tumors of the ovary dysgerminoma and the Brenner tumor

The *granulosa cell tumor* is a malignant feminizing hormone producing tumor It causes precocious sexual development in girls and rejuvenation in older women These tumors may occur at any age but are commonest in childhood They may attain a large size filling the abdomen Generally they are from 4 to 5 inches (10 2 to 12 7 cm) in diameter The most constant feature of the histologic structure is its variability which can be identified only by an expert pathologist

A precocious sexual development is characteristic The sexual changes are more noticeable before puberty and after the menopause than at intermediate ages In childhood granulosa cell tumors cause precocious development of the reproductive organs and menstruation begins in a normal fashion but at much too early an age

After the menopause a granulosa cell tumor causes sexual rejuvenation The breasts become full the vagina and external genitalia resume the features of mature sexual life The uterus returns to its normal size and menstruation is resumed It may be irregular or periodically normal If therefore hyperplastic endometrium is obtained on biopsy in the search for the cause of postmenopausal bleeding a granulosa cell tumor is present if the patient has not received estrogen therapy

Carcinoma of the uterine tube occurs both primarily and secondarily Primary carcinoma of the tube is rare secondary carcinoma is common in abdominal and pelvic carcinomatosis

Carcinoma is usually situated in the middle or outer third of the tube The gross appearance of the tumor depends on its size and the situation of and patency of the fimbriated extremity of the tube If the fimbriated extremity is open peritoneal implantation is the rule leading to ascites carcinomatosis and death Metastasis and dissemination occur early and are widespread

The first symptom of carcinoma of the uterine tube usually is pain Pain is followed by a vaginal discharge which is bloody The first symptom may be abdominal enlargement due to ascites and peritoneal carcinomatosis

The pain may be dull with sharp exacerbations It may be as severe as the pain in a tubal abortion but without shock because rarely is there any internal hemorrhage in tubal cancer and hence there are no syncope and shock Menstrual irregularities of all types occur the commonest being metrorrhagia and menorrhagia

Secondary carcinoma of the uterine tube is commoner than primary carcinoma Being usually metastatic it generally first starts in the tubal wall as discrete nests and then extends into the tubal lumen

The diagnosis of carcinoma of the uterine tube is made from histologic studies

THE OVARIES

The ovary lies longitudinally or obliquely against the outer wall of the pelvis From its upper end proceeds the suspensory or infundibulopelvic ligament and from its lower end the utero ovarian ligament The normal graafian follicles and corpus luteum can be palpated on the surface of the ovary in thin women

Congenital Malformations Absence of one ovary is rare but when it occurs the absence is due to complete agenesis of one urogenital fold and the corresponding kidney is also absent In the absence of one urogenital fold in the female the uterus is unicornous derived from only one mullerian duct The uterine body is fused with the single uterine tube The other uterine tube is likewise absent

In the ovaries when there is a follicular agenesis there is no follicular epithelium The uterine tubes uterus and breasts are underdeveloped The axillary and pubic hair is absent or scanty and the external genitalia are infantile in type The urinary gonadotropins are elevated as they are after menopause The 17 ketosteroids are low Often there are many other and severe congenital and acquired anomalies The absence of genital development is often the least of the disabilities present as the result of congenital malformations In these patients the ovarian failure is a basic lesion in the ovary itself

Tumors Tumors of the ovaries are often complex structures histologically and may defy the knowledge and ingenuity of the pathologist to classify them They may be benign or malignant or they may consist of immature male gonadal elements which secrete male hormonal substances which produce masculine secondary sex characteristics Grossly on examination by physical means they are found to be either cystic or solid in structure benign or malignant in nature Whether or not they are cystic or solid does not separate them structurally into benign and malignant groups for both cystic and solid tumors of the ovaries may be found to be malignant

Retention Cysts These cysts are benign but may become malignant and originate serous cystomas and cystadenomas Retention cysts arise from occlusions of germinal epithelium The germinal layer of epithelium may be present only in the fissures that separate lobulations of the ovary Cysts are formed when this germinal epithelium is occluded by adhesion

Follicular Cysts The follicular cysts may originate in normal ovaries or as the result of an inflammatory thickening of the ovarian capsule which prevents rupture of the follicle Follicular cysts are 1⅕ or 1⅗ inches (3 or 4 cm) in diameter and occur bilaterally

Of the secondary carcinomas of the ovary the *Krukenberg tumor* (fibrosarcoma ovarii mucocellulare carcinomatodes) involves both ovaries and is usually preceded by or accompanied by a neoplasm in the stomach intestinal tract or breast As a rule there is a history of a previous neoplasm in one of these regions The original focus of the growth may not have been suspected however and it is not discovered until the pelvic tumor is discovered Krukenberg tumors usually occur in women between the ages of 20 and 60 years

Not all secondary ovarian carcinomas have their primary origin in the alimentary tract For instance carcinoma of the breast may metastasize to the ovary At least 7 of each 10 of the secondary ovarian tumors are of the same cell type as the original growth

The symptoms of secondary ovarian carcinoma may be those of the primary growth in the stomach intestine or breast or may be referred to the pelvis or to an enlarged abdomen On pelvic examination there are bilateral solid ovarian tumors Often there is ascites Bilaterally enlarged ovarian tumors in a woman who has a tumor of the breast stomach or intestines are diagnostic The original tumor may have to be sought for after the bilateral ovarian tumors are found The prognosis is generally hopeless

Tumors Arising in the Ovum (Teratomas) Teratomas are embryonic tumors which are characterized by containing one or all of the three primordial layers of tissues ectoderm mesoderm and entoderm Teratomas are organized to varying degrees into special organs such as hair teeth skin bone cartilage thyroid intestine and other organs

The younger the ovum is when the growth disturbance takes place the more nearly totipotential its cells are and the more capable of producing all their tissues The older the ovum the more specialized the cells and the less capable they are of producing a variety of tissues and thus the origin of dermoid cysts (MacCallum)

Dermoid cysts are simple forms of teratomas Teratomas and dermoids may be found anywhere in the body but have a predilection for the ovary

Malignancy is common in teratomas in dermoids it is relatively rare

Struma Ovarii The ovarian tumors which contain thyroid tissue are termed struma ovarii These are in reality ovarian teratomas

Struma ovarii is distinguished by its ability to produce symptoms of hyperthyroidism before removal and occasionally after removal of the thyroid gland The tumor has a high iodine content This tumor is aberrant endocrine tissue and hence secretes hormones

The chief symptom is an enlarged and uncomfortable lower part of the abdomen At times there may be mild abdominal pain Much of the abdominal enlargement is due to the ascites which is often caused by peritoneal implantations of the tumor On examination there may be the signs and symptoms of hyperthyroidism accompanied by ovarian enlargement

When hyperthyroidism is present in association with an ovarian tumor struma ovarii is likely present Hyperthyroidism may persist after thyroidectomy The removal of an ovarian struma may be followed by recovery or at times hypothyroidism and myxedema Generally the prognosis is excellent unless the tumor is malignant

Connective Tissue Tumors of the Ovary Fibromas and sarcomas are rare tumors of the ovaries They may occur at any age although sarcomas are found most commonly in younger persons even in children fibromas in adult women These tumors may attain 4 to 6 inches (10 2 to 15 2 cm) or more in diameter They occur bilaterally and are often associated with ascites and hydrothorax

Fibromas differ from sarcomas by remaining within the limits of the ovary There are varieties in appearance and color of fibromas and sarcomas but the characteristic gross difference in these tumors is that sarcomas are much more vascular than

On palpation the tumor is smooth and may be firm often it contains cystic portions but usually it gives the impression of a solid mass

The diagnosis of a granulosa cell tumor is established by the presence of an ovarian mass sexual precocity or rejuvenation and the histologic presence of cells suggesting or resembling granulosa cells of primordial follicles of the ovary

In children the other causes of sexual precocity are cortical adrenal hyperplasia or tumor pituitary tumors and disorders of the hypothalamus The removal of a cortical adrenal tumor is followed by disappearance of precocious sexual development The temporary sexual precocity does not seem to have any permanent ill effect

The *arrhenoblastoma* is a malignant ovarian tumor associated with the defeminization or masculinization of a woman It is to be differentiated from adrenal cortical hyperplasia or tumor of an adrenal gland Arrhenoblastoma is always to be considered as a malignant tumor but as in the malignant granulosa cell tumor recurrence may be very slow in developing

At first there occurs a slow process of defeminization of the woman Then follows the development of masculine traits such as hypertrichosis growth of beard increase in length of clitoris deepening of the voice and development of masculine contours

Adrenal tumors of the ovary are malignant tumors and are said to be histologically like cortical adrenal tissue resembling the hypernephroma type of cell The histologic biologic and clinical resemblance between cortical tumors of the adrenal gland and arrhenoblastomas indicates an embryologic kinship

Cortical adrenal tumors masculinize females and cause sexual precocity in boys In all cases of manifest sexual changes precocity sex reversal or rejuvenation the ovaries adrenals pituitary and hypothalamic disorders (see Chapter 14) are studied

The *ovarian dysgerminoma* is usually malignant It does not exert any influence on sexual development or the hormonal balance It is the homologue of the seminoma an embryonal carcinoma a common testicular tumor The dysgerminoma is supposed to arise in rests of the undifferentiated mesodermal cells of the primitive ovary before these cells develop either male or female characteristics This tumor has a tendency to occur in patients who have sexual maldevelopment such as hypogenitalism and hermaphroditism

Dysgerminomas may be rather large tumors and often bilateral They are hard elastic at times nodular and may be accompanied with ascites Some of these tumors are malignant others are possibly benign

The *Brenner tumor* is considered to be a benign tumor It is a rare growth which secretes no hormone When of small size it appears as a solid mass but when large it may appear as a cystic adenoma with large nodular masses on its surface The Brenner tumor is diagnosable only by histologic study

Carcinomas of the Ovaries Ovarian carcinomas occur as both solid and cystic tumors The solid carcinoma of the ovary develops directly from ovarian tissue The cystic carcinomas develop from some other ovarian tumor such as a papillary cystadenoma Epithelial tumors develop in the dermoids and the teratomas of the ovary

Solid carcinomas of the ovary begin on the surface of the ovary involve the whole gland and extend to involve surrounding structures the broad ligaments pelvic floor and peritoneum They metastasize freely especially to the lungs They do not cause any early symptoms The patients first complain of abdominal enlargement which is due mainly to ascites

On examination carcinoma of the ovary is difficult to palpate even when fairly well developed In a woman in whom bimanual examination is difficult an insensitive ovarian carcinoma is often impossible to feel

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fibromas In sarcomas secondary growths in the pelvis or throughout the *peritoneal* cavity on the intestine and in the omentum may be present The *pelvis may contain* bloody fluid if implantations are present

Sarcomas are among the most malignant of ovarian tumors Because they rarely produce any symptoms until they are well developed cures are rare Metastasis and direct invasion with peritoneal implantation *are the common modes of extension*

The diagnosis of both fibromas and sarcomas is made by microscopic study of the tissue The presence of a solid insensitive freely movable hard mass in the region of the ovary suggests a *fibroma* If ascites is present the diagnosis is more *nearly certain The diagnosis of sarcoma* of the ovary is more difficult than that of fibroma because the tumor is softer and may be confused with other ovarian tumors

Complications of Ovarian Tumor *Twisting or torsion of the pedicle* of an ovarian tumor is associated more commonly with dermoid cysts fibromas and cystomas than with other types of tumor If the rotation occurs suddenly it instantly produces unbearable pain nausea vomiting and shock When the twisting develops slowly there are often recurring attacks of moderate pain which may become more severe and constant as the twist becomes tighter

Examination in the case of twisted ovarian pedicle reveals an abdomen that is acutely tender and in the later stages rigid The ovarian mass on pelvic examination usually is palpable The final diagnosis may not be made until laparotomy is performed

Rupture of ovarian cysts is common and occurs in cysts that have thin distended walls There is usually a history of a fall There may be abdominal discomfort and often there is severe pain In an occasional instance a cyst may rupture and drain through the vagina without serious consequence

Free perforation is common in endometrial and corpus luteum cysts Peritoneal irritation or peritonitis develops if the contents of the cyst are chemically irritating or infected

Intra abdominal hemorrhage may follow the rupture of an ovarian cyst and produce severe symptoms very similar to those of rupture of ectopic pregnancy Hemorrhage of varying degrees may occur in connection with ovulation and the rupture of a normal graafian follicle and this tends to recur with subsequent ovulation

CHILDBIRTH INJURIES

Mechanical Injuries Mechanical injuries which occur as the result of pregnancy or childbirth commonly involve the reproductive organs the urinary organs and their supports and the abdominal wall Mechanical injuries which involve the supporting structures in the back or the bony pelvis are less frequent

Mechanical injuries of the reproductive organs and the supporting structures result in lacerations of the perineum in varying degrees (first degree second degree and third degree) or complete perineal tear rectocele enterocele cystocele relaxation of vesical sphincter descensus and retrodisplacement of uterus prolapse of uterus cervical laceration and infection prolapsus of vagina inversion of uterus and rectovaginal vesicovaginal and ureterovaginal fistulas Some of these injuries such as relaxation of the vesical sphincter can be discovered only by special examination (endoscopic examination) although they may produce characteristic symptoms

Rectocele is a hernia of the rectum into the vagina It is due to stretching or tearing of the rectovaginal septum Abdominal and rectal pressure causes the rectum to bulge into the vagina through the weakened rectovaginal septum

Enterocele is located in the recto uterine pouch and is a hernia through the pelvic floor It may contain small or large intestine omentum or the a

Cystocele is a protrusion or partial hernia of the bladder due to a stretching and relaxation of the vesicovaginal septum The intra abdominal pressure and the weight of the bladder and intestine cause the bulging of the bladder into the vagina

Descensus of the uterus can occur only when the broad ligaments and utero sacral ligaments are sufficiently relaxed to allow it Uterine descensus may be of various degrees of severity its end stage is prolapse

Prolapses of the uterus are of three degrees In *first degree* prolapse the cervix descends to the ischial spines in a *second degree* prolapse the cervix appears at the vaginal orifice in a *third degree* prolapse the whole uterus is extruded from the vagina Third degree prolapse and procidentia are synonymous

Prolapse of the vagina occurs in women in whom the vaginal wall lacks adequate support or in whom the supporting structures were so frail that prolapse was inevitable

Vaginal fistulas are vesicovaginal ureterovaginal vesicocervicovaginal urethrovaginal or other combinations The commonest type is the vesicovaginal with urinary incontinence All such fistulas are rare

Injuries to the Cervix Many parous cervices show unhealed areas round the external os infection and lacerations which may be only partly healed The lacerated and infected cervix may be big boggy and heavy and thus may increase the weight of the pelvic organs An unhealthy cervix produces annoying vaginal discharges dysmenorrhea pelvic pressure backache and urinary frequency

Injuries to the Uterus The commonest endometrial disorder from childbirth is an endometritis as the result of puerperal infection or retained placental tissue Such an endometritis may become chronic The commonest permanent change in the uterus however is chronic subinvolution

Injuries to the Urinary Organs A discomfort in the pelvis or in either iliac fossa may be due to a urinary lesion probably in the ureters or the renal pelvis as a consequence of parturition Urologic examinations are required for confirmation of urologic disease to account for the postpartum urinary symptoms

Injuries to the Abdominal Wall and the Pelvis The *abdominal wall* is stretched during pregnancy and there may remain diastasis recti and hernia—femoral inguinal or umbilical—which were created or made worse by gestation

The coccyx is frequently traumatized a condition which is manifested by local symptoms

The *supporting structures* of the pelvis may be injured by a strain of the ligamentous structures Some of these injuries are the result of the unnatural postures assumed for instance the extreme lordosis from marked increase in the abdominal weight

Symptoms of Childbirth Injuries A sense of pressure in the pelvis and a feeling as though the pelvic organs were protruding into the vagina while the patient is standing are common complaints Backache fatigability nervousness irritability lack of strength and energy and constipation are not necessarily manifestations of birth injuries of the pelvis

Leukorrhea and urinary frequency are common symptoms of a childbirth injury In cystocele and prolapse there is a partial hernia of the bladder which prevents complete emptying of the bladder by urination and gives rise to subsequent infection and cystitis Urinary symptoms vary from frequency that accompanies cystocele and prolapse to partial incontinence due to an injury of the vesical sphincter Partial incontinence manifested by the loss of a few drops of urine on laughing heartily coughing sneezing quickly flexing the body or lifting is a very common complaint

Complete incontinence is the result of a vesical fistula or destruction of urinary control A constant dribbling of urine while standing in a woman who can hold the urine while lying down and can empty the bladder normally when it is filled often indicates a ureterovaginal fistula

Anal incontinence may be due to a third degree tear, even though it was repaired at the time or may be due to a rectovaginal fistula

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The period of maximal fertility in women is between 18 and 30 years of age Men may retain their fertility much longer than women

The menses need not be regular in order for the ovaries to produce normal ova If the menstrual periods occur although in time they may occur irregularly and if there are present cyclic changes in the cervical mucosa it generally can be assumed that ovulation is proceeding normally If there can be demonstrated changes in body temperature during the menstrual cycle excretion of pregnanediol in the urine changes in the hormone content of the blood and urine and cyclic changes in the vaginal mucosa ovulation is assuredly proceeding normally

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Local deformities or disease or both in the genital organs constitute the commonest causes of sterility These local conditions may exist in the vagina vulva cervix uterine body uterine tubes and ovaries

The *normal vaginal secretions* are so acid that they kill spermatozoa very quickly Hence the only spermatozoa that are likely to reach the uterine cavity are those that are deposited on or near the cervix The *cervical secretion* is alkaline and exhibits cyclic changes in amount cellular content alkalinity viscosity and permeability between the ninth and nineteenth days of the cycle

Dyspareunia may be a factor in sterility only when it prevents delivery of the sperm into the vagina It may be due to a variety of lesions imperforate hymen rigid hymen narrow vaginal orifice following infections spasm operative or obstetric scar absence of vagina or other congenital defect In the absence of the vagina the urethra may be injured in attempts to use it as a copulative organ

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volume however, vary considerably and volumes from less than 2 ml to more than 7 ml may be normal Very small amounts of semen may mean occlusion of the spermatic ducts

Normally the *semen is thick and viscid* when ejaculated It should liquefy readily however and be fluid within less than half an hour At least one half of the *spermatozoa should be actively motile* after 2 to 10 hours

Brown has made some investigations of male seminal fluid with a view to determining whether or not modification in the character of this fluid or increase in its amount might serve to protect the spermatozoa over a longer period of time and make more likely the contact of the sperm cell with the ovum Man normally produces about 120 000 000 spermatozoa for each milliliter of fluid or approximately 450 000 000 in the average ejaculate A donor may produce a total of 30 000 000 to 50 000 000 spermatozoa every 24 hours daily over a considerable period When it is realized that only one spermatozoon can impregnate the ovum the tremendous factor of safety involved becomes apparent These studies lead to the conclusion that such influence as the glandular secretions of the male have on the spermatozoa cannot be further increased by the addition of fluid to the normal ejaculate

If more than 1 in every 5 spermatozoa are found to be abnormal it is evidence of impaired fertility

A *decrease in the normal number of spermatozoa* is one of the commonest causes of low fertility Spermatozoa counts of less than 60 000 000 per milliliter are inadequate

Absence of spermatozoa in the semen results either from failure to produce sperm (aspermia) or from mechanical blockage of the seminal ducts Congenital abnormalities of these structures are hypospadias absence of the vasa deferentia epididymides seminal vesicles or ejaculatory ducts imperfect union of testis and epididymis and congenital stricture of the vas deferens

Lack of spermatogenesis may be due to the various types of pituitary disease or *to destruction of the germinal epithelium of the testis from inflammatory* lesions such as orchitis of mumps or tuberculosis or to failure of normal testicular development as in cryptorchids

Functional Impotence Functional impotence of the male may be due to inability to have an erection because of fear of the sexual act fear of having children or apprehension that he may be unable to perform the sexual act normally Worry of any sort may abolish normal sexual power

It is well for the diagnostician to remember that many men and women who after various tests and examinations have been declared sterile nevertheless have enjoyed rearing their own families despite the grave predictions to the contrary

DERANGEMENTS OF SEX LIFE

The consultation is a solemn moment to the patient man or woman Under its influence patients find themselves confessing to the physician and asking for help which might well be sought from a minister of the gospel When a patient is in this mood the physician can well listen and by listening he is practicing sound morality and sound psychology It is just as much the physician s affair to mend the patient s body by helping him to forego sexual irregularities as to encourage him to stop drinking himself to death Sexual irregularities are never normal sexual behavior

Description of what should be considered a normal sex life can only lead to hopeless disagreement not only with one s peers but even with one s self It is not the prerogative of the physician to outline the normal sex life to men and women for the sex impulse is a normal appetite as variable as any that can be imagined The physician is never afraid to hold up to his patients the frank ideals of a cleaner life than it would seem possible for some of them to aspire to The strictest morality that can be imagined must always be outlined as routine behavior In time delight

will come to those who give such advice for often a responsive glow a determination to be better will be aroused in the most unpromising material

Morally upright and profoundly religious mothers may have read certain books about The Things Women Should Know About Their Sex Life If such a woman has not experienced an orgasm she may inquire of the physician the reason for her lack of such an experience

In answer to such a query it should be briefly but clearly stated that in women orgasm is a nervous reaction based on the presence of healthy reproductive organs and psychic response It is dependent too on normal pituitary and ovarian function If desire and orgasm have been developed without frustration they continue irrespective of the presence of the ovaries or uterus Even though these reactions have not been attained if normal genital organs are present reproduction may be possible for there are many women who have never experienced an orgasm who have large families It is therefore clear that while the orgasm is desirable it is not necessary for breeding in women For the physician to enter into a psychiatric cross-examination of such a woman is nothing short of bestial brutality for such questioning is sure to offend her finer sensibilities

The removal of the ovaries shortens the span of sex life by altering the physical character of the external genitalia and the vagina Women become indifferent sexually and have less orgasm after menopause

SEXUAL OFFENCES

The physician is consulted in regard to two types of sexual offence The first is a mental disturbance of which the crime is only a manifestation These crimes consist of repeated indecent exposure indecent assaults upon children lewd and libidinous practices and the criminal acts of sadism and masochism

The second group are deliberate sexual crimes by persons in a sane though perhaps morally degraded frame of mind Rape incest and sodomy and bestiality are the common offences

Rape Rape is unlawful carnal knowledge of a woman without her consent Unlawful implies that there is such an act as a lawful carnal knowledge of a woman by force Carnal knowledge is the slightest degree of penetration of the vulva by the penis Penetration proper through the hymen is not necessary to the act When the hymen is already deflorate ruptured by previous penetrations a great deal depends on the girl s character and bearing the substance of her story the condition of her clothing and the presence of even trivial injury to any part of the body

Evidence of a forceful assault freshly committed will be clear There will be bruising scratching or tearing of the vulva or hymen which may still be bleeding

It is possible that the only proof of penetration will be a vaginal smear of seminal fluid For this reason a swab or smears should always be taken The emission of seminal fluid is not essential to the proof of carnal knowledge but its demonstration may provide proof of penetration

The development of gonorrhea or of pregnancy in a woman after alleged rape may sometimes be used as corroborative evidence without giving it undue value and culture as well as microscopic examination of a swab should be made if possible

The essence of the offence is that it should take place against the desire of the woman In females less than 18 years of age the offence is carnal abuse with or without consent The age limit of 18 years may vary in accordance with local statutory law

Medical Examination in Alleged Rape The precise time of the examination and a record of by whom it was requested are essential to record

The raped girl or woman is shy and bewildered The story in the girl s own words is recorded at the time for it will contain conversation the exact form of which may be vital No false modesty in detail or language should be allowed to interfere with this being a full record It will gain by being graphic and tend to appear in its true light—authentic or false

The presence of tangled disordered hair dirt stained face or hands and the condition of the clothing tears and blood stains are recorded

With some person present to assist the clothing should be removed in the doctor s presence and examination made of every part of the body Stories of being forced against something pinned down gripped by the arms kicked or punched may gain corroboration from the finding of some comparable injury in the part described

The local parts are examined for foreign hairs to be placed in envelopes sealed and labeled Seminal stains are smeared on slides and fixed Bruises scratches or tears adjacent to the vulva and within it as well as to the neighborhood of the hymen are searched for The state of the hymen is observed and recorded

Matted semen stained pubic hairs may be cut away Dried thigh stains should be moistened with saline solution and smeared on glass slides A vaginal swab should always be taken If the material is still wet the peculiar odor of semen competently observed is fairly conclusive

It is less common for the doctor to have the opportunity of examining the man accused of rape permission must be obtained and its voluntary nature made clear before commencing The physician searches for foreign hairs on the clothing adherent to the body or pinned under the prepuce Injury to the penis is rare but scratches to the face or hands occur frequently (Keith Simpson)

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8

DISEASES OF THE SKIN, SUBCUTANEOUS TISSUES AND BREASTS

The skin is a protective respiratory excretory heat regulatory absorptive vitamin producing and immunity producing organ Its prickle cell layer contains the sense organs for tactile pain heat and cold sensations

Layers of the Skin The skin is composed of two main layers or parts (1) the epidermis or cuticle and (2) the true skin The *epidermis* or cuticle (scarf skin) is composed of the following layers beginning from the outside the horny layer (stratum corneum) the transparent layer (stratum lucidum) the layer of flattened granular cells (stratum granulosum) the mucous layer (stratum mucosum) which contains pigment and prickle cells (prickle cell layer) the basal cell layer (stratum germinativum) The *true skin* consists of an outer or papillary layer which contains the nerve endings and capillaries and a deeper layer which is largely composed of fibrous bundles Beneath the skin is the subcutaneous connective tissue

The expanse of skin over the body is designated by topographic regions for instance skin of the scalp and skin of the leg These are topographic regions which normally grow hair The regions which do not grow the regular coarse hair are designated as glabrous skin

Pigmentation of the Skin Eyes and Hair The stratum mucosum of the epidermis as has been stated contains the pigment or coloring substance the melanin The melanin is composed of small granules which are deposited in the deep layers of the epidermis and iris and disseminated through the cells of the shafts of the hair The mucous membranes sclerotics and some other tissues of the body contain this substance The pigmentation of an individual is subject to change throughout different periods of life Other changes are brought about by age and exposure to sunlight and to confinement The state of feeling of the individual and the presence or absence of an oily coating of the skin likewise influence its color In the hair the presence of air bubbles between the hair cells and the loss of melanin cause grayness of the hair normally with aging and allegedly suddenly after great emotional strain

The function of the pigmentation is to protect the individual against sunlight The absence of pigment results in albinism with its accompanying sensitive white skin and reddened eyes which are extremely sensitive to light

Racial Colorations Early man was a product of the tropics and subtropical regions He had brown skin hazel to dark brown eyes and reddish brown to black hair Coon believes that man reached Europe about the middle of the Glacial period During the early part of the post Glacial period he penetrated into northern Europe and the Scandinavian peninsula

Because of the climatic influences of the post Glacial period the original skin pigment began to be lost and as a result of these environmental influences and mutations the skin became white and blond coloration came into being Along with the white skin came blue eyes and hair ranging from light brown yellow and golden to almost white In the central zone of Europe there was less depigmentation than in northern Europe and thus the Celto Slavic or Alpine coloration came into existence while in southern Europe depigmentation was still less leaving much of the original skin coloration and resulting in the Mediterranean complexion of swarthy

brown with dark eyes and black hair The continuous residence of those remaining in the tropical countries seems to have increased their skin coloration and thus resulted in the black skinned individuals (Negroes) The Nordic white skin the blue eyes and the blond hair still contain some of the ancient pigment The eyes are blue because this old brown pigment is limited to the posterior cells of the iris and the blue coloration results from refraction, whereas when the iris is viewed from behind it is brown or dark

Freckles (*Ephelis Lentigo*) The familiar freckles of the skin occur in those who have red hair or a tendency thereto also in individuals who have white skins or in admixtures of white skinned and black skinned individuals

A freckle is a pigmented spot brownish in color on the skin It is due to discrete accumulation of pigment in the rete Freckles vary in size in color and in number on different individuals They are made prominent by exposure to sunlight and wind The term cold freckles is applied to those occurring on a part of the body under the clothing which is not exposed directly to the sun The origin of these freckles is the same as of those caused by direct exposure to the sun They are less densely pigmented spots

Freckled skins are proved to be susceptible to senile keratosis and skin cancer in aging individuals

Leukoderma Leukoderma occurs at all ages but among white peoples it is more common in the middle aged than in the young The eruption commences as a small white patch which may enlarge and spread The disease is more frequent and extensive in hot climates and during the summer months than in other climates and seasons The lesions often occur on the face hands scalp and trunk and may be symmetrically distributed The individual patches do not reveal thickening of the skin and when they are examined for the sense of touch and pain anesthesia is not present When the scalp is involved the hair on the patches turns white

The normal skin sensitivity is of importance in the differential diagnosis between leukoderma and leprosy Scleroderma may show some whitening of the areas involved but has a distinct boardy hardness Some observers incline to the view that leukoderma is of nervous origin Actinic rays in many cases have an influence in determining its origin and localization Differentiation of leukoderma from the fully developed lesion of pinta is difficult In pinta however there will be other lesions of varying colors which have not yet become white (For more about pinta see Chapter 17)

Chloasma Chloasma is a disease the opposite of leukoderma There is a spotted increase in the normal color of the skin In chloasma the increased pigmentation has an internal origin Chloasma (or chloasma like spots) is associated with pregnancy There are many cases of uncertain origin Chloasma is erroneously attributed to hepatic disease (liver spots)

Acanthosis Nigricans Acanthosis nigricans is a rare disease affecting the flexures and often resembling ichthyosis The brown and at times the almost black velvety linear overgrowth in the region of the flexures is characteristic The mucous membranes often are affected showing warty growths without pigmentation This condition frequently is associated with widespread internal malignant disease Besides resembling ichthyosis acanthosis nigricans resembles Darier s disease

Transient Coloration Texture and Moisture No decision concerning the color of the skin should be made until the mucous membranes of the mouth and conjunctivae have been observed Exposure to wind and weather dries hardens and darkens the skin as well as producing other effects of less obvious magnitude *Habitual pallor* is seen in persons who live an indoor life or who sleep in the day time A transient pallor of the skin may be the result of weakness of the heart action as is observed in syncope chills nausea and arterial spasm Sudden pallor which is constant for the time being is present when large and rapid hemorrhages

occur and when the heart fails abruptly A slowly developing pallor is usually the result of anemia The pallors are often described in terms of varying tints In diagnosis however such descriptions are worthless

An unusual redness of the skin depends on hyperemia the overfilling of the cutaneous capillaries with blood Hyperemia of the skin follows a warm bath friction of the surface of the skin or exercise Long exposure to heat and cold and moisture produces a purplish redness of the skin of the hands which is not significant of systemic disease *Diffuse redness* of the skin is seen in fevers after the administration of certain drugs and after the ingestion of alcoholic beverages Localized redness of the skin such as the redness of one cheek in acute lobar pneumonia and the unilateral redness of the face which attends histamine cephalalgia (Horton s headache) are observable phenomena In some *chronic alcoholics* the face is a dusky red Not all persons who have dusky red faces however are chronic alcoholics The so-called dusky red face is the result of cyanosis of the skin which is characterized by a blue or purplish tint This color arises from imperfectly oxygenated blood in the capillaries *Cyanosis* is best observed in the lips and mucous membranes and in the fingernails if nail polish has not been used because of the thinness and translucency of these epithelial coverings Cyanosis of the skin is caused by any condition which hinders the admission of air to the lungs lessening the working breathing surface of the lungs and by any condition which interferes with pulmonary or systemic circulation Stated in more precise terms cyanosis is due to improper oxygenation of the erythrocytes The *texture of the skin* is soft and elastic and the skin is moist to the touch

HYPERHIDROSIS AND ANHIDROSIS Excessive moisture or sweating of the skin is *hyperhidrosis* and dryness of the skin is *anhidrosis* (see Autonomic Nervous System Chapter 23)

Sweating of the hands or the feet attends general debility hyperthyroidism and Raynaud s disease It is most commonly due to nervousness Unilateral or one sided sweating of the head and face may be present in histamine cephalalgia (Horton s headache) neuralgia and other affections of the nervous system Unilateral or bilateral sweating of the face may occur during a meal

Anhidrosis is characteristic of all febrile diseases especially if the temperature is high and the rise is prolonged If the skin is injured by stretching which alters its structure so that the cutaneous circulation is hindered as in general anasarca lymphedema or myxedema anhidrosis exists as a direct result Anhidrosis accompanies diseases in which there is an excessive loss of fluids for any reason When renal functional activity is severely impaired the sweat may have a urinous odor and may deposit white scales or crystals of urinary solids on the skin

COLORED SWEAT Yellow sweat from the biliary pigment may occasionally be present in severe jaundice Blue brown yellow or red sweat has been observed in hysteria The condition characterized by colored sweat is known as *chromhidrosis* Very rarely bloody sweat *hematidrosis* is caused by capillary hemorrhage into the sudoriparous glands This condition may follow the administration of dicumarol

Dermographia Dermographia is a condition in which tracings made on the skin leave a distinct reddish elevated mark *Dermographia alba* is the white line formed on the skin in the epinephrine test *Black dermographia* is the dark streak produced under certain conditions by stroking the skin with certain metals

According to Urbach and Pillsbury the black writing which appears on the skin following application of certain blunt metallic implements with moderate pressure represents not a chemical but a physical process due to the particles of the metal rubbed off by friction and remaining on the skin While it was originally assumed that a hysterical condition was a necessary prerequisite for the phenomenon Emdin later retracted this idea

Black dermographia is frequently observed in women who wear trinkets and

cheap jewelry of silver and gold on powdered areas The same effect occurs on skin surfaces soiled with street factory or mine dust

In principle the phenomenon may be evoked on any human skin by the use of appropriate inorganic powders There are individual differences due to variations in the moistness and oiliness of the skin The phenomenon will appear in all persons who have dry nonoily epidermis The fact that the phenomenon can be evoked in the skin of a corpse conclusively demonstrates that this process cannot be regarded as a biologic reaction

History When a disorder of the skin is present it is well to learn (1) the duration of the disease (2) whether there have been previous similar attacks of the eruption (3) whether the onset of the disease was accompanied by fever malaise headache or generalized pains or has been accompanied by disturbances of digestion or of the urogenital or nervous systems and (4) whether there have been any enlarged lymph nodes

Examination The examination of the skin should always be conducted in daylight if at all possible Of basic importance in diagnosis are detailed examination of the lesion or lesions especially of the primary lesion and palpation of the eruption Primary skin lesions are known as macules papules vesicles pustules wheals and hives The secondary skin lesions are described as scales crusts scabs excoriations fissures ulcers and scars These primary and secondary lesions may be acute or chronic The primary lesions are of the greatest importance in diagnosis A magnifying glass used to distinguish the character of the lesion may be of great aid The examination is performed in a definite order beginning with the head mouth neck and proceeding to the thorax upper extremities lower extremities and finally the soles of the feet and the palms of the hands It is during the examination that the examiner often is able to obtain a precise history as to whether the eruption began universally or was widely scattered or was grouped on the flexor or extensor surfaces of the body During the time of the examination the patient usually will mention whether the lesions began by itching burning hyperesthesia anesthesia or with pain

There are predispositions to many diseases of the skin During childhood the common predisposing causes of skin disease are first those of hereditary origin for instance the congenital nevi and nevoid diseases which include such disorders as ichthyosis and keratosis palmaris Congenital syphilis may not become manifest until adolescence In later life there is a tendency toward seborrheic eczema presenile alopecia and pigmentary disorders of the skin

Infants are subject to various infantile dermatoses and to napkin eruptions Acne seborrhea and hyperhidrosis occur in adolescence whereas in advanced age epithelioma keratosis and general pruritus develop The relation of sex to skin disease is often important Carbuncles occur twice as often in men as in women Women are more subject than men to lupus erythematosus Leukomelanoderma of syphilis is practically limited to women Women suffer from contact dermatitis from the use of cosmetics whereas men suffer more from occupational dermatitis and leukoplakia of the mouth During the winter months such diseases as psoriasis seborrheic dermatitis ichthyosis and chronic eczema are worse than in summer Scabies and contact dermatitis due to soap wool and furs are commoner during the winter than in summer During the summer months diseases of the skin as the result of contact with various poisonous plants hyperhidrosis urticaria dermatomycoses and miliaria are commoner than during the winter In summer the insects that sting or bite and the animal parasites that suck blood are prevalent

DISEASES OF THE SKIN

DISEASES DUE TO PRENATAL INFLUENCES

Ichthyosis Ichthyosis is both familial and congenital It has been thought to be due to a persistence of a fetal layer of skin the epitrichium

In cases of ichthyosis the secreting glands of the skin are absent Perspiration is impossible and the patient suffers in hot weather from vascular congestion and the lack of oil In cold weather the extremities are susceptible to chilblains The skin is easily irritated is hard horny in appearance with marked dryness harshness and formation of heavy scales Only the flexures escape involvement The hard masses of scales often crack across and in so doing produce fissures which become infected with pyogenic organisms

The mildest phase of ichthyosis is termed xeroderma All gradations between xeroderma and typical ichthyosis are observed In the completely developed ichthyosis large platelike scales are present all over the body but generally are worst on the extremities

Keratosis Palmaris et Plantaris This is a hereditary and congenital disease of the skin which affects the palms and soles and often is manifest at an early age The lesions in appearance are like keratotic lesions on senile skin

Lichen Pilaris (Keratosis Pilaris) Around the hair follicles along the anterior surfaces of the thighs and over the outer surfaces of the arms are often observed small firm papules that are not inflammatory These papules are discrete and at times may itch They are arranged in patches and are ill defined and frequently some of them are capped with crusts of dried blood as the result of scratching Keratosis pilaris usually does not produce symptoms

Epidermolysis Bullosa This is often a hereditary condition which begins in infancy and is manifested by bullae which appear after the slightest trauma In the development of the lesions there is first redness then wheals and the bullae appear in less than one half hour The patient s skin is more elastic than normal Often there is dystrophy of the nails Epidermal cysts are of common occurrence

Adenoma Sebaceum This is a rare condition which even more rarely occurs alone It is often associated with congenital nevi fibromas warts rhabdomyoma and convulsive disorders and defective minds

The lesions are manifested soon after birth as small flattened tumors of the sebaceous glands on the face In the beginning the yellowish individual lesions are small 1 or 2 mm in diameter Around these yellowish areas there often are dilated capillaries

Keratosis Follicularis (Darier s Disease) Morbid heredity is the only known cause of Darier s disease which runs in families and commences in youth There are no lesions of the mucous membrane The disorder is not associated with malignant disease Although the disease normally is chronic and slowly progressive acute exacerbations may occur with the rapid production of fresh patches but the general health of the patient is not impaired

This disorder is characterized by crusted dark brown papules often coalescing in symmetric areas on the face scalp trunk flexor aspects of the limbs axillae and genital regions When the crusts are removed from the papules there are discernible on the undersurface of the crusts horny plugs corresponding to dilated follicular openings Removal of the crusts causes bleeding Sebaceous material can be squeezed out of the follicles after the crusts are removed

Monilethrix Monilethrix is a congenital and familial dystrophic condition in which the hair shows regular nodal constrictions at which it is likely to break off Associated with the dystrophy is keratosis pilaris a hyperkeratosis of the follicular stomas The hair is thinly distributed in the beginning After many hairs have broken off the remaining hairs are set beside the short ones both long and short hairs display variations in thickness and impart a shabby appearance to the scalp

Hereditary Xanthomatosis Bloom and his co workers recorded hereditary xanthomatosis as it occurred in 9 children of Syrian descent whose parents were second cousins The parents of these tainted siblings had no cutaneous or cardio vascular symptoms although the father had hypercholesteremia The mother had

8 half siblings, and 5 of them also had hypercholesteremia This appears to support the hypothesis that hypercholesteremia rather than xanthoma tuberosum is the principal directly inherited factor and that this disturbance of lipid metabolism under certain conditions leads to cutaneous and cardiovascular changes The hereditary cause of the disorder has been demonstrated but no adequate statistical data are available that warrant a final analysis of the genetic mechanism involved (see Lipidosis Chapter 22)

Xanthomas are small flat plaques of a yellow color in the skin Microscopically the lesions reveal light cells with foamy protoplasm

Tuberous Sclerosis Tuberous sclerosis is a rare heredofamilial disease in which there are multiple neurospongioblastomas in the brain The nodules and the accom panying sebaceous nodules of the skin have a moderately firm consistency which is fancied to be that of a potato hence the name tuberous Tumors often coexist in other organs notably hypernephroma of the kidney and rhabdomyoma of the heart The child is usually epileptic and mentally deficient

The presence of sebaceous adenomas small closely set papular tumors of the skin pink or brownish appearing symmetrically on the face chiefly about the nasolabial folds bridge of the nose forehead and chin is the chief diagnostic feature The nodules sometimes are on the skin of the trunk and the intergluteal fold These cutaneous growths appear in adolescence and therefore the condition may be difficult to recognize prior to this age The tubers may be situated within the cranial cavity and originate the localized or generalized symptoms of brain tumor

BACTERIAL INFECTIONS OF THE SKIN

Infections by the staphylococcus are the most common infections of the skin The staphylococcus produces furuncles impetigo contagiosa pyoderma sycosis vulgaris and pyogenic paronychia The streptococcus too is a common cause of skin infections It causes more serious infections of the skin than the staphylococcus such as ecthyma erysipelas and scarlet fever

Staphylococcal and Streptococcal Infections *Acute staphylococcal derma tosis* is characterized by pustular follicular lesions The appearance of the lesions varies with their depth The commonest form of the dermatosis is a superficial infection of the openings of the hair follicles These lesions are common and give little or no discomfort Usually they are brushed away by bathing or by the clothing without one s knowing of their presence Deeper infections of the follicles result in boils or furuncles

Furunculosis is more frequent in hot weather than at other times and may result from the rubbing of clothing such as a collar suspenders belt or a cuff at the wrist if there has been a previous lesion pus from it may remain on the clothing to be rubbed in somewhere else

A furuncle usually is caused by a staphylococcal infection in a hair follicle The lesion begins singly about the neck under the arm or in some other hairy portion of the body and is first a central small pustule surrounded by inflammatory indura tion Almost from the beginning the lesion is painful and tender The furuncle inspired the original classical description of an acute localized infection namely rubor tumor calor and dolor Boils or furuncles destroy the follicles and on healing leave scars A furuncle is distinguished from a carbuncle by the fact that a furuncle has only one opening whereas a carbuncle has multiple openings

Metastatic infection in bones kidneys and other distant points is not a rare complication of furunculosis it occurs however in a relatively small proportion of cases It is probable that pressure squeezing incision or other trauma to furuncles produces a transient bacteremia which in turn results in metastatic abscesses (see the Fevers Chapter 17)

Carbuncles often are caused by streptococcal infection They are much larger than furuncles are painful and often cause fever and leukocytosis Frequently they occur on the back of the neck and occasionally on the upper lip Owing to the peculiarities of the lymph drainage of the upper lip and the lower part of the nose, furuncles and carbuncles situated here are particularly dangerous because extension of the infection may enter the subdural spaces almost unhampered Both furuncles and carbuncles may be the initial foci of infection which is spread by the blood stream to the heart lungs pleura peritoneum meninges and bones

Hidrosadenitis is the term sometimes applied to axillary and inguinal affections by furuncles The axillary lesions do not always suppurate and often are of the type called blind boils The affection may extend to all of the axillary skin and sub cutaneous tissues There may be difficulty in examining the area properly owing to the difficulty and pain caused in attempts to abduct the arm from the side

The prognosis in all the acute skin lesions caused by the staphylococcus and in some infections due to the streptococcus depends on understanding the normal progress of the lesions The defensive cellular reactions initiated in the skin and subcutaneous tissues lead not only to the formation of pus but to an effective barrier or wall around the region of infection which prevents its extension The collection of pus is shut in by this protective wall occupies the center of the necrosed follicle and forms the core of the boil The lesion should be allowed to burst at its weakest point or at most the weakest point lanced and the contents and core allowed to evacuate spontaneously a process which leaves a more or less aseptic cavity Healing quickly ensues if the barrier has not been cut through by a knife or pierced deeply with a lancet At most superficial lancing is all that should be permitted

Sycosis vulgaris is a chronic superficial folliculitis affecting mainly the beard The infection may be most intractable It is maintained not by chronicity of the individual lesions but by their constant repetition The disorder may persist for years with little or no loss of hair indicating that the hair roots have not been involved However too much emphasis should not be placed on the superficiality of the lesion for at times epilation of the hairs of infected follicles is not painful indicating that the follicle is affected Owing to the chronicity of the infection there are general redness and thickening of the affected region of the skin The presence of a chronic pustular inflammation of the hair follicles limited to a hair bearing area especially the beard is often sycosis vulgaris

The three common affections of the beard are impetigo which clears quickly tinea more chronic and sycosis vulgaris which often lasts for years

Seborrheic sycosis or sycosiform dermatitis is not limited like sycosis vulgaris to the beard The beard of men and the eyebrows and eyelashes of both men and women may be affected Affections of the eyebrows and eyelashes are common The infection begins after the age of puberty and produces painful swollen red eyelids and eyebrows The eyes matter and often are difficult to open on arising in the morning

On examination a chronic blepharitis with loss of eyelashes is present The blepharitis may appear after the dermatitis or may never be noticed There is a patchy dermatosis a mixture of folliculitis with red scaly dermatitis spreading beyond the hair margin Similar patches occur on the upper lip in the grooves of the lower lip and chin and under the chin A distinct hypertrophy of the skin in the affected areas may be present These patches are often persistent lasting for years without a change in size

Granuloma pyogenicum is staphylococcal in origin An injury has allowed the staphylococci to enter The lesions are on the fingers at the nail fold They are small red shiny pedunculated swellings composed of granulation tissue which bleeds easily

Streptococcal dermatitis is a superficial inflammation of the skin characterized by an acute onset of redness thickening of the skin, and an abundant exudation of serum In either the localized or the generalized forms of the disease the folds of the body especially those behind the ears and the scalp are more profoundly and frequently affected than other regions of the skin As the inflammation subsides there is formation of fine lamellar scales

Erysipelas begins at one point on the skin of the face or neck and spreads rapidly with a well defined edge from this point The lesion is inflamed hot tense shiny and painful Secondarily occurring over this patch may be vesicles If vesicles do not form there is no subsequent scaling of the skin Erysipelas is always a serious disease Its prognosis is bad in the aged and the debilitated (see Chapter 17)

Impetigo contagiosa in some instances seems to be due to the streptococcus and in others to the staphylococcus The vesicle if cultured in the early stage may yield streptococci or staphylococci It may be argued that the staphylococcus is either the primary or the secondary invader Children are most frequently affected since infection is spread from clothing and contact Isolation of these patients should be mandatory

Superficial areas of the horny and the malpighian layers of the skin are the only parts involved At first there are vesicles filled with clear fluid The fluid becomes cloudy and the vesicle ruptures Crusts form which are composed of dead epithelium inspissated serum leukocytes fibrin and cocci When these crusts are shed the papillae are not exposed and there is no bleeding Healing is rapid and complete

Examination reveals yellow crusts and vesicles situated mainly around the mouth the cheeks ears and occasionally on other parts of the face In the more severe infections the scalp and the fingers are involved Often there are pinkish macules indicating areas where the crusts have been shed from the face and scalp Pediculosis capitis may initiate the scalp infection and cause the spread and continuance of sporadic cases of the disease

When the fingers alone are affected the lesions spread peripherally and for this reason may resemble a similar type of lesion on the face (*impetigo circinata*) which looks like ringworm The reason for this resemblance is that healing of the bulla begins in and proceeds from the center At the periphery the infection remains active spreads and gives rise to a crusted circular lesion which increases in size

Impetigo neonatorum (pemphigus neonatorum) is a streptococcal infection which occurs in infants and is characterized by bullae about the mouth and finger nails The disease is not pemphigus

Impetigo is highly infectious Not infrequently epidemics of the disease break out in lying in hospitals The disease is often contracted by the mother On her the bullae appear about the breasts or places of contact All precautions are employed to prevent dissemination of the infection such as thoroughly washing and sterilizing clothing dishes drinking cups and feeding bottles All who attend these patients wash their hands thoroughly after the examination before handling any objects

The diagnosis is based on the appearance on examination and on a recent occurrence of the disease in the institution or neighborhood The disorder is differentiated from chicken pox pemphigus dermatitis herpetiformis dermatitis or eczema and tinea circinata

Impetigo barbae is a refractory form of impetigo which spreads rapidly and proves refractory to treatment This disorder is limited to the bearded regions and is characterized by the ease of epilation followed by a drop of pus The disease is contracted in unsanitary barber shops

Infective intertriginous dermatitis or a similar disease may be caused by streptococci or other organisms The commonest sites of this disease are the folds behind the ears, with or without extension to the scalp Parts of the horny layer of the

skin are removed irregularly leaving a raw area The fold is thus rendered red and wet while the scalp may be dry and scaly Despite careful handling when the ear is moved forward to permit inspection of the fold the thickened swollen epidermis may crack and bleed The disease is characteristically chronic and will return until all foci in the scalp have been eradicated

Acute infective dermatitis may result from various organisms The lesions are asymmetric superficial well defined erythematous vesicular pustular scaly patches The margins of the lesion are undermined the horny layer is raised as a scale with its free edge toward the center of the lesion It may not be possible to culture the causative organism

Chronic dermatitis is common There are three important conditions which occur in association with chronic dermatitis namely varicose veins injuries to skin nerves or blood vessels and discharging wounds

Varicose dermatitis is a common type of infective dermatitis In varicose dermatitis the inner surface of the lower third of the legs is affected As the disease progresses ulceration develops which leads when healed if healing takes place to the formation of rigid bound-down scars These scars may encircle the limbs by a hard fixed cuff of skin and subcutaneous tissue which further impedes the venous circulation

A dermatitis encircling a long discharging wound or sinus often occurs The dermatitis is severe if digestive chyme is discharged from the sinus

Dermatitis may follow injury to the skin of the feet hands or other exposed parts particularly if the injury has required long bandaging and dressing with salves The injury may be from any cause but often there has been a puncture wound or injury to the underlying tissue as well as the skin A dermatitis which is difficult to heal may attend injury of a trunk of a peripheral nerve

Infectious eczematoid dermatitis is distinguished by pustular and impetiginous lesions perhaps due to the staphylococcus but more frequently to several and usually indeterminate organisms Inappropriate treatment often irritates this sort of dermatitis On examination there is actually a contact dermatitis present

The appearance of the lesions of eczematoid dermatitis especially the margins patch tests and microscopic examination of the marginal scales prepared with sodium hydroxide for fungus will help to limit the etiologic possibilities Often the lesions will clear up if treatment other than cleanliness is discontinued

Acne conglobata is characterized by hypertrophy of the skin which accompanies the lesions of acne This hypertrophy is most noticeable about the nose on the neck back and buttocks These hypertrophic skin lesions of acne may become granulomatous and may form freely discharging sinuses often deep and extensive They may partially heal On each reactivation of the draining sinuses a generation of furuncles may occur These sinuses may resemble those of a tuberculous infection or one of the mycoses such as sporotrichosis In a patient of middle age a rapid increase in the area of induration around the sinus without other evidences for a secondary infection arouses the suspicion of a superimposed malignant degeneration

Diphtheria Diphtheria may be complicated by a severe ulceration of the skin The ulcers have membranous bases and reddened cyanotic appearing margins These lesions may persist for weeks or months after perhaps a mild diphtheria

When a diphtheritic ulceration is suspected smears and cultures from the lesions are taken at once and if diphtheroid bacilli are found antitoxin should be administered Virulence tests of the organisms may be made in order to arrive at a definite diagnosis

Tuberculosis Tuberculosis of the skin is a serious but fortunately a rare disease Three women suffer from the disease for each man so affected The disease is one of youth before the age of 20 years It is acquired through contact by an extension of an internal infection or by blood borne infection

The tuberculous lesions on or about the nose and face are contracted by the direct spattering on the skin from coughing or sneezing of saliva or mucus which contains tubercle bacilli The postmortem wart occurring on the hands of pathologists is also an example of infection from contact Butchers too may contract similar lesions from infected cattle

The skin over infected and caseating lymph nodes and around the sinus from tuberculous osteomyelitis may be infected

Tuberculous septicemia may cause skin lesions over the body as a part of the miliary process Miliary tuberculosis of the skin often appears suddenly during the course of pulmonary or bone tuberculosis and during convalescence from one of the exanthemas such as measles

Tuberculosis of the skin except for miliary tuberculosis is a chronic disease The lesions progress by centrifugal spread There are central healing and scarring while the periphery is active and slowly spreading The patient who has tuberculosis of the skin may die at an old age with the disease still active if there are no other active foci present in the body When other active foci such as an active pulmonary tuberculosis or tuberculosis of the bone are present the prognosis is that of the original infection The skin lesion may clear up and leave the other foci active

For the purposes of description tuberculosis of the skin is usually divided into three forms namely (1) lupus vulgaris (2) scrofuloderma and (3) miliary tuberculosis

Lupus vulgaris or tuberculosis cutis luposa is characterized by a great degree of chronicity It seems to be an inoculation type of tuberculosis which occurs on the head face nose upper lip cheeks ears and neck of children from 4 years to 10 years of age The lesions are thus distributed because it is obtained from small droplets of tuberculous sputum disseminated in the air by coughing or sneezes of a tuberculous individual

The process and the subsequent scarring may cause contracture of the mouth destruction of the soft parts of the nose with necrosis of the cartilage leaving a flat gaping orifice to the posterior nasal cavity ectropion and perhaps blindness through corneal irritation and subsequent scarring or loss of the auricles by destruction of the soft parts and subsequent necrosis of cartilage

The usual lesion is a nodule from 2 to 5 mm in diameter It is raised or it may lie on a level even with the surface In consistency it is softer than the surrounding skin so that a blunt point drawn across the nodular area sinks as if crossing a hole in the skin In color the lesion is a light brown clear and almost transparent The characteristics of the lesion are better observed if a glass slide is pressed firmly over it in order to expel the blood from the part The nodules often coalesce so that a larger area of affected skin appears which is actually one lesion There is a tendency to heal behind the spread of the lesion and healing leaves irregular scars The mucous membranes frequently are involved

Examinations of the lungs roentgenologically and of the urine and the sputum microscopically and by means of guinea pig inoculation for tubercle bacilli are important diagnostically A positive skin reaction to tuberculin seldom is helpful if the reaction is negative it rules out tuberculosis The diagnosis depends on a skin biopsy However in a young person the demonstration of the clear brown or reddish chronic nodules characteristically distributed on the face nose and lips accompanied with scarring and secondary epithelial changes leading to crusted warty forms is usually sufficient for a diagnosis of lupus vulgaris The prognosis is bad The disease persists throughout life

Scrofuloderma (scrofula) is a direct infection of the skin from tuberculous lymph nodes of the neck or other underlying tuberculous infection Draining sinuses develop and the adjacent skin is inflamed swollen and cyanotic Syphilis for practical purposes is ruled out if the serum reaction is negative

Miliary tuberculosis of the skin is but a part of the disseminated tuberculosis It is a terminal manifestation of the disease elsewhere Small papules appear which soften and ulcerate and if life is prolonged the papules spread and coalesce and form ulcers These lesions may occur about the mucous orifices without general distribution as a result of direct inoculation from infected discharges The diagnosis is obvious

SYPHILIS OF THE SKIN

Syphilis Syphilis was formerly an important cause of skin disease In tropical climates syphilis and yaws are still the cause of much suffering

Congenital Syphilis Congenital lesions of syphilis become manifest during the first year after birth The important lesions of congenital syphilis are enumerated as (1) interstitial keratitis (2) Hutchinson s teeth and (3) labyrinthine disease (see Diseases of the Lens Chapter 4)

The skin lesions consist of a wrinkled loose appearing skin which has been described as old mannish in appearance A pemphigoid or papular eruption may be present on the palms and soles and over the legs The bullae of the pemphigoid eruption are of longer duration than those of impetigo neonatorum The papular lesions are commoner in the anal and the genital regions than on other parts of the body These papules may be erythematous in appearance and may closely resemble those of acquired syphilis

Often neither parent of a congenitally syphilitic child will admit knowledge of a syphilitic infection The physician is in turn interrogated concerning two possible sources of the child s infection (1) Could it have come from the child s grand parents? (2) Could syphilis have been accidentally acquired? The answers are (1) Congenital syphilis is from the mother to the child (2) Accidental infection occurs but usually syphilis is acquired as are all venereal infections and that is by direct contact which is usually through sexual intercourse However accidental inoculation may occur through a breach in the skin or in a mucous membrane by such means as infected vessels for eating and drinking infected instruments especially the tattoo needle kissing infected persons droplet infection of a physician s eye during examination of a syphilitic throat or direct infection of a finger during the examination of a patient In some instances an admission of these means of acquiring syphilis may save much time for the physician However in reality the physician should realize that it may not be too important how syphilis was acquired if the patient has it

Syphilis characteristically and specifically affects the arteries producing endarteritis As a consequence of endarteritis there is an exudation into the tissues Palpation reveals that as the result of the endarteritis and exudation into the true skin the skin is thickened and inelastic

Acquired Syphilis The *primary lesion* (hard chancre) appears from 2 to 6 weeks after infection If no ulceration has occurred there is a hard raised red papule the surface of which is glazed moist and bleeds on being traumatized If the primary sore is on the external genitalia the patient always ascribes it to trauma during intercourse If ulceration has occurred there is an eroded red papule which soon becomes indurated and later circumscribed raised moist hard with an inflamed surrounding area of skin or mucous membrane The chancre usually lasts from 2 to 8 weeks No scar is left if there has been no ulceration Scars are permanent after ulceration The regional lymph nodes are enlarged early The primary lesion may be small may last only a week or two and is described as just a pinpoint sore

The diagnosis is confirmed by the presence of the treponema in serum from the lesion on examination under darkfield illumination Positive results of serologic tests are not present for several weeks

Extragenital chancres occur around the fingernails the face, lips cheeks mouth breasts abdomen and in fresh tattoo injuries of the skin

The *secondary lesions* of syphilis are general surface lesions and skin eruptions In many instances the skin lesions of syphilis may be precisely described as macular papular and nodular syphilides

Symptoms referable to the body as a whole or general symptoms vary in degree and are not predictable There are often a feeling of ineptitude pains through the body affecting the arms and legs and headache which occurs at night after the patient has retired to bed There may be a low grade fever during the evening hours In an occasional instance there are high fever and skin eruption which spreads rapidly causing a serious disability This is the so called malignant syphilis These rapidly developing secondary lesions of syphilis occur over the body and involve the mucous membranes as well Involvement of the mucous membrane by syphilis may be severe with ulceration and deep sloughing of the tonsils and nasopharynx

A syphilitic eruption usually commences as a macular erythematous eruption over the abdomen trunk and face The eruption of syphilis is commonly present on the palms of the hands and on the soles of the feet In the beginning it is truly a macular eruption There is no itching The eruption varies in color and intensity and may simulate that of measles and of influenza The color of a syphilitic eruption has often been described as coppery or fawn The lesions are in not on the skin

The *macular syphilid* is a common macular lesion that occurs on the body This lesion often commences around the umbilicus and from this part spreads over the thorax and the flexor surfaces of the extremities, it occasionally extends to the neck but rarely to the face The average diameter of the lesions is about 1 cm The edges are ill defined The lesions are at first a light yellow which changes to a reddish color There is no itching and no scaling Usually there are other symptoms of syphilis present such as the scar of the initial lesion or chancre There also usually are enlarged lymph nodes in the posterior triangle of the neck and in the axillary epitrochlear and inguinal regions Alopecia may be present Sore throat and general aches and pains may be present and in women there may be a history of miscarriage

There are two forms of *papular syphilid* One form the small papular variety usually occurs over the flexor surfaces of the extremities As a rule the lesions are grouped The papules are small and discrete may be slightly scaly and sometimes are associated with pustules On palpation the lesions can be felt situated in the *corium The second form the large papular variety may occur* on the face neck and genitals as flat papules often clearing up in the center thus sometimes being designated as an annular type of syphilid These lesions are sometimes scaly almost *always accompanied by induration and usually by other symptoms* of syphilis

The *nodular syphilides* in syphilis are late secondary or tertiary lesions These nodules may begin as millimeter sized papules which are well defined Frequently they are multiple two or more in number The individual syphilid is firm and smooth and reddish in color As the disease advances the nodules increase in size ulcerate and have the so called punched out appearance that is characteristic of syphilitic ulcers These lesions often accompany other symptoms of syphilis They are painless

During the secondary stage of syphilis the serologic reactions are positive

A large papular eruption of syphilitic skin lesions may closely simulate lichen planus Lichen planus is itchy and occurs on the palmar surfaces of the wrists the inner aspects of the knees and the legs below the knees

Both psoriatic lesions and syphilitic lesions may be circinate Psoriasis is a disease of long standing and recurrent manifestations whereas syphilis is a disease of recent manifestations and rapidly spreading lesions and involves the true skin

Leukoderma frequently occurs about the region of the neck and is accompanied

with pigmentation around the characteristic small white areas Stubborn patches of pinta and syphilis about the face and neck often lasting for years simulate the lesions of chronic psoriasis or tuberculosis A unilateral psoriatic appearing lesion on the buttock calf or sole should be regarded with suspicion The ringed and horseshoe manifestations of syphilis may resemble psoriasis or ringworm but neither of these diseases affects the true skin

The serologic tests for syphilis are now most reliable and diagnostically dependable However there is a possibility of false positive serologic reactions In all questionable instances of these reactions a decision about validity of the test usually can be made from the history and the findings All false positive and serologic fast reactions should be checked by examination of the cerebrospinal fluid

INFECTIONS OF THE SKIN DUE TO VIRUSES

Eczema Herpeticum Kaposi s varicelliform eruption and Juliusberg s pustulosis vacciniformis acuta are synonymous names for eczema herpeticum

Eczema herpeticum is due to the herpes simplex virus and occurs in children who as a rule already have an eczema In the majority of cases according to Barton and Brunsting this eczema is a primary herpetic infection and as such is accompanied by a severe systemic illness which occasionally is fatal A less severe form of the disease is observed in those who have circulating antibodies in the blood In these patients there is often a history of contact with a manifest infection by herpes simplex

The close association with atopic eczema may implicate the virus potentiating activity of histamine or the wide expanse of denuded skin so common in these patients may merely provide a good nidus for exogenous infection with the virus The disease may resemble eczema vaccinatum

The vesicles develop rapidly in large numbers and in successive generations for 3 or 4 days to a week The largest number of the varicella like vesicles occur on the already eczematous skin A small number of grouped lesions appear on the intact skin Those which appear first undergo desiccation rupture and expose the corium or they become encrusted and shed The outcome in the majority of cases is favorable

The diagnosis can only be suspected clinically and is established with certainty by demonstration of the virus by viral technics

Varicella Herpes Viruses The similarities of varicella and herpes zoster lie in the nature of their viral agent (*Briareus varicellae*) their vesicular eruptions and their intranuclear inclusion bodies while their greatest differences are found in age incidence epidemiology and changes in the central nervous system hence their widely different symptomatology (see Viral Infections Chapter 17)

Varicella or chickenpox sometimes also called herpes zoster zona shingles or zoster is a mild communicable disease caused by a virus similar to that which induces herpes zoster *Herpes zoster* is an affection limited to the cutaneous distribution of a cutaneous nerve

Varicella (*Chickenpox*) The incubation period is usually from 2 to 3 weeks In 24 hours after the onset of fever papules of irregular distribution followed by or accompanied with vesicles with surrounding erythema appear on the face and trunk spreading in some cases to the mouth and pharynx and extremities In chickenpox the lesions are usually most abundant over the trunk as contrasted with the face and extremities in smallpox The eruption occurs simultaneously with the fever and its duration is proportional to the height and persistence of the fever Successive generations of lesions appear The vesicles may not become papular and may seem to be set on the skin However they frequently are papular and usually become pustular and the crusts are often removed by scratching because of itching Impetigo furuncles septicemia and glomerulonephritis may complicate the disease

Vesicles on the laryngeal mucosa about the eyes and genitalia and in the hair may become infected secondarily and cause much discomfort

On occasion an encephalitis, symptomatically similar to that accompanying herpes zoster and measles complicates chickenpox Patients usually recover from chickenpox encephalitis

Verrucae (Warts) Warts are small benign papillary overgrowths which vary in size color and shape They occur slightly more commonly in women than in men Certain families seem to have a relatively greater susceptibility to infection than other families Warts are auto inoculable and transmissible and may disappear without treatment There are four distinct types according to Lane verruca vulgaris verruca plana juvenilis verruca plantaris and verruca acuminata

The abnormal cellular hypertrophy is due to a local infection with a virus Whether separate viruses cause the four different forms of verruca or whether *the different forms are clinical variations resulting from one virus is unknown* Rudloff expressed the belief that venereal vegetations and verruca vulgaris are sister growths due to the same filtrable virus which manifests itself by papilloma formation on mucous membranes or on thin skin and by wart formation elsewhere

Verruca vulgaris is the commonest type These warts occur predominantly in children and adolescents and appear most often on the exposed areas particularly on the hands and fingers but any portion of the skin surface may be involved Variants of this type are the threadlike filiform wart of the eyelids and face and the digitate warts which project from the scalp face and neck

The lesion of verruca vulgaris is a raised circumscribed solid round gray or yellowish gray papule varying in size from 2 to 10 mm The surface at first is flat and smooth but may become scaly crusted or fissured The warts may be single or multiple frequently there is a primary mother wart which in time is joined by satellite lesions

Verrucae planae juveniles known as flat warts are slightly raised small flesh colored round or angular papular lesions which are distributed over the face neck hands and wrists Almost universally multiple these lesions occur most often in children and young women They may resemble vesicles or may closely resemble the papules of lichen planus and rarely those of molluscum contagiosum They produce a dirty appearance on the face when they have coalesced and are situated on the chin

Verrucae of the plantar type occur on the padded areas of the soles and heels less frequently the sides of the feet the under surfaces of the toes the palms and palmar surfaces of the fingers There may be a single wart but often there are small satellites in the vicinity of the original lesion Differentiation from a callus and a clavus may be difficult but is facilitated by paring the surface of the verruca A *wart is revealed as a well defined discrete gray round or oval area* often containing small dark specks or threadlike lines which are deposits of blood pigment If the surface of a callus or corn is pared a smooth hard yellow growth containing no blood stains is seen

A variant of the plantar wart is the so called *mosaic wart* which has been well described by Montgomery and Montgomery as a warty patch horny and granular with an irregular border and varying in width from 1 to 10 cm Comparatively painless the patch is situated under the pressure points and may extent onto the toes and behind the heels It is often mistaken for callus Paring reveals the intra cellular *nature of the patch which consists of many* small individual cores so closely grouped that they resemble a mosaic pattern The pain and tenderness of plantar warts may be sufficient to hamper the activities of the patient seriously

Verruca acuminata known also as condyloma acuminata and venereal wart occurs chiefly on or about the genitalia and the anus in the axillae and groins and on the navel As single or multiple discrete or confluent raised *filiform or*

papillomatous lesions whose surfaces are often moist and macerated from the secretions of the surrounding organs they are attached by broad or narrow bases Verrucae acuminatae may be sensitive painful and malodorous and may interfere with function

Warts may be difficult to treat successfully Many patients are convinced of the therapeutic virtues of witchcraft for the cure of warts

INFECTIONS OF THE SKIN DUE TO HIGHER PLANT ORGANISMS
(*The Superficial Mycoses*)

Fungi microscopic members of the plant kingdom belong to phylum Thallophyta orders Algae and Fungaceae The order Fungaceae comprises bacteria (Schizomycetes) true fungi (Eumycetes) and slime molds (Myxomycetes) The Myxomycetes are nonpathogenic Of the four subclasses of true fungi the Hyphomycetes (Fungi imperfecti) contain the common pathogenic fungi present in lesions of the skin and hair A skin fungus which does not invade the hair follicle is termed *Epidermophyton* Sabauraud called the fungi which proliferate on the surface of the hair Microspora the two groups (Trichophyta) which invade the hairs are endothrix and ectothrix depending on the situation of the spores in relation to the shaft of the hair

Ringworm Ringworm infection of the skin according to Lewis and Hopper is arbitrarily divided into regional varieties namely scalp (tinea capitis) beard (tinea barbae) smooth or glabrous skin (tinea glabrosa) folds of skin (tinea cruris) and feet and nails (dermatophytosis and onychomycosis)

Tinea Capitis (*Ringworm of the Scalp*) Tinea of the scalp usually is due to one of two Microspora *Microsporum audouini* and *Microsporum lanosum*

However both the *Trichophyton* and the *Microsporum* group have (1) species which are pathogenic principally for certain lower animals (zoophilic) and (2) species which are pathogenic principally for human beings (anthropophilic)

Infections contracted from animals have two important characteristics they are but slightly if at all contagious among human beings and they either are self-limited or respond to topical therapy Humans are poor hosts for this group of fungi and as Lewis and Hopper showed the self limited features of these fungi may be accounted for by the development of antibodies and the production of an inflammatory reaction in the neighborhood of the infected hairs

In the United States *Microsporum lanosum* is the important animal (zoophilic) representative of the Microsporum group and is generally transmitted by cats or dogs Unlike the disease contracted from cattle *Microsporum lanosum* usually produces but slight inflammatory reaction Several members of a family may be infected from the same pet but extensive epidemics do not occur Infections produced by *Microsporum lanosum* must be differentiated from the epidemic form (*Microsporum audouini*) because they are self limited and nonepidemic

Infections with human (anthropophilic) fungi are contagious and are usually resistant to topical therapy The lack of antibodies and severe inflammatory reaction is ideal for survival of the fungus and allows infection to persist for years

The genus *Trichophyton* has several species in the anthropophilic group called endothrix because the spores invade the shaft of the hair Infections with endothrix *Trichophyton* are rare and nonepidemic in the United States

Microsporum audouini has been responsible for the recent rapid increase in ringworm of the scalp in the United States

Microsporum audouini may cause epidemics in institutional children When the condition is first discovered there are several small areas in the occipital and temporal regions in which the hair is dull and broken off The surface of the patch is usually scaly Occasionally a considerable degree of inflammation may be present but this is not the rule The lesions spread peripherally finally reaching 1½ inches (3 cm) or

more in diameter There is little tendency for the infection to spread to other parts of the body

Trichophyton violaceum develops insidiously and is widely dispersed over the scalp It invades the cortex of the hair causing the hair to break off close to the scalp or just below the surface of the scalp which gives the appearance of an ingrowing hair The disease may heal at puberty

Microsporum lanosum causes a fungous infection of the scalp of children and occasionally involves glabrous skin It may be at times contracted from pets (kittens) The initial lesion is frequently the largest There is loss and breaking off of hair in the affected patches with marked inflammation and tenderness The duration of the disease is only a few months

Microsporum fulvum resembles *Microsporum lanosum* except there is a tendency for the inflammation to remain localized to only one part of the scalp

Endothrix *Trichophyton* causes favus Favus usually is a follicular involvement of the hairs but it may occur with minimal follicular involvement The disorder may spread to the nails or to the glabrous skin

On the scalp favus begins as a small area of scaly inflammation in which the hair follicles become festered Breaking of these small pus pockets results in yellow crusts which increase in size and finally become cup shaped (scutula) The scutula are yellow friable scabs pierced with hair There is intense itching and a moldy odor like that of rats The hair of the scutula falls out The skin becomes atrophic and due to the spread of the disease an irregular permanent baldness ensues Spontaneous recovery from the disease eventually occurs However when there is a minimal follicular involvement there is only a diffuse superficial but adherent scaling of the scalp with little if any permanent alopecia

Trichophyton gypseum is often contracted from pets and farm animals It may vary in intensity and distribution of its manifestation in accordance with susceptibility to it Widespread infections involving the hands feet and face are due to increased susceptibility or increase in the virulence of the infective agent The characteristic lesion is a severe inflammatory reaction of the scalp and the development of kerion

In many instances bacterial infections of the skin and fungous infections are both present This is particularly true of an infection like *Trichophyton crateriformi* When a mixed infection is present part of the manifestations—the crusting kerion and shedding of hair—is caused by the secondary infection by bacteria and other fungi The treatment of the bacterial infection often suffices to effect a recovery

In summary the manifestations of tinea of the scalp are partial loss of hair in patches breaking off and lack of luster of the infected hair and varying degrees of inflammation Atrophy and scarring may follow certain types of infection Kerion formation is common In appearance a kerion is a marked tissue reaction to the infecting microorganism one of several species of fungi The pustular lesion is a painful elevated boggy erythematous localized tumefaction with simple scaling at the bottom

DIAGNOSIS The diagnosis of ringworm of the scalp may be made reasonably accurately by microscopic study of scrapings from the lesions skin or hairs when properly prepared

Patients who have ringworm of the scalp vary in their reaction to trichophytin mainly in accordance with the type of infecting microorganism and with their own powers of resistance There is usually more response when the fungus is also pathogenic to animals

A scalp infected with ringworm will show under filtered ultraviolet rays a characteristic fluorescent effect whenever infected hair is present The ultraviolet rays are diagnostically important in revealing the presence and extent of the ringworm infection of the scalp

Tinea Barbae (*Ringworm Infection of the Beard*) Infections of the beard commonly are due to *Trichophyton gypseum* There are occasional instances of infections caused by *Microsporum lanosum* *Trichophyton rosaceum* *Trichophyton violaceum* and *Trichophyton purpureum* The spread of these infections is thought

to occur in the barber s shop Animals are known to be carriers of *Trichophyton gypseum* or *Microsporum lanosum* The disease may be contracted by contact with those who have infections of the scalp or of the glabrous skin There are two types of infection (1) diffuse inflammatory and (2) sycosis

The diffuse inflammatory process is contracted from pet animals The beard in the affected areas loosens and epilation is easy In the sycosis type of tinea barbae once the infection is established it slowly spreads as a mild crusted folliculitis with breaking off of the invaded hair (*Trichophyton violaceum*) or similarly without the formation of hair stumps (*Trichophyton purpureum*)

The diagnostic value of the reaction to trichophytin depends on the type of infecting organism *Trichophyton gypseum* and *Microsporum lanosum* infections give definite positive reactions to skin tests In the other infections the skin reactions are irregular and cannot be depended on as tests of diagnostic value

If the shaft of the hair has been invaded a fluorescence of the infected hair is observed under filtered ultraviolet radiation Endothrix organisms give the fluorescence The ectothrix organisms produce no fluorescence under the filtered ultraviolet radiation The inflammatory type of tinea barbae has to be differentiated from sycosis barbae iododerma or bromoderma and syphilis

Tinea Glabrosa (*Ringworm of the Smooth Skin*)_ Fungous infections of the glabrous skin occur as scaly circinate patches as solid plaques or in geographic configurations

Microsporum lanosum (*M canis*) obtained from contact with kittens or other pets or from an infected individual is the usual cause of ringworm of the smooth skin Less commonly infection of the smooth skin extends or is carried from scalp infections (endothrix *Trichophyton*) or from feet infections (*Trichophyton gypseum Trichophyton purpureum*)

Microsporum lanosum infection is manifested by an erythematous ringed lesion which may gradually increase to a diameter of 5 or 6 inches There are usually minute vesicles along the border the surface is scaly and the center appears unaffected *Sometimes two or more concentric rings may appear in a single lesion* The lesions are commonly present on the faces or necks of children with tinea capitis The backs of the hands and other exposed parts are the usual sites of the first lesions in patients who catch the infection from an outside source New lesions may develop on various parts of the skin depending on extension and on transfer of the organisms from the original focus The subjective symptoms are usually mild although the infection is itchy and subsequent scratching may contribute to its spread

The eczematous type of smooth skin infection by tinea may simulate in appearance almost any form of dermatitis The primary eczematous type is the initial response to infection (usually *Trichophyton gypseum*) In this form of eczematous dermatitis the first lesion (or lesions) is the vesicle and vesicles continue to form rupture and become crusted The infection spreads peripherally there being no tendency to central clearing The lesions remain ill defined or become circinate with active vesiculation along the periphery

The secondary type of eczematous dermatitis from tinea is caused by medicaments or by treatment producing primary irritation which transforms the circinate lesions into vesicular oozing patches surrounded by varying degrees of dermatitis

Trichophyton purpureum may cause a large surface eruption of skin of the trunk The infection begins at one or more points and migrates in a thin line over an ever widening area The affected skin is dull red and presents a slight infiltration and scaling on the surface As the border of the lesion advances behind it the skin is lighter owing to loss of color Persistent itching is a constant symptom

The diagnosis is usually established by cultural methods from the skin lesions

Fungi are absent in material taken from the lesions The trichophytin reaction is usually negative in cases of uncomplicated tinea of the glabrous skin

The prognosis varies with the infecting microorganism Ordinary tinea circinata usually responds to treatment within a week or two The eczematous type takes longer to respond endothrix *Trichophyton* infections still longer and the infections due to *Trichophyton purpureum* are resistant

Tinea Cruris (*Dhobie Itch*) Tinea cruris is a superficial fungous infection (caused chiefly by *Epidermophyton floccosum* and *species of Trichophyton*) confined usually to the medial surface of the proximal third of the thighs although other parts of the skin may become affected The infection may be obtained from contact or from infected articles of clothing Another form of the disease is a contact dermatitis of various causes in India it has been attributed to the marking fluid bhilawanol oil (from *Semecarpus anacardium* of the family Anacardiaceae) used on laundry by the native washermen (dhobie) Tinea cruris at times may assume *epidemic heights*

The eruption is usually bilateral and symmetric it favors the upper inner portions of the thighs but may extend up to the pubis and as far back as the sacrum The axillae the umbilicus the inframammary areas and the interdigital webs of the feet are occasional sites of the infection Vesicopustules are sometimes seen on the soles The friction of clothing during hot weather causes varying degrees of secondary eczematization

The *area of the rash is well marginated by a border of minute* vesicopustules The surface is scaly and there is little or no tendency to central clearing The lesions are brownish red

The absence of evidence of tinea infection of nails hair feet or smooth skin elsewhere over the body limits the likelihood of other tinea infection

Tinea cruris usually does not initiate sensitization to trichophytin A test with trichophytin usually elicits a negative or a mildly positive reaction With *Trichophyton gypseum* a positive reaction is usual but when *Trichophyton purpureum* is the causative fungus a negative reaction is common

The ordinary form of the disease usually responds readily to medicinal applications but tends to relapse The lesions caused by *Trichophyton gypseum* are likewise readily cured but those due to *Trichophyton purpureum* are extremely obstinate

Dermatophytosis The term dermatophytosis refers to a superficial fungous infection of the skin The disease (sometimes called athlete s foot) is usually acquired by transfer from organisms affecting the hands feet or nails

Dermatophytosis is a common disease It is commonest in boys and men from 15 to 25 years of age Hyperhidrosis is commonly present in those who have dermatophytosis

Some lesions of the feet and nails resemble dermatophytosis of fungous origin but may be due to causes other than fungous infections If the dermatophytosis is due to a fungus *Trichophyton gypseum* and *Trichophyton purpureum* can be cultured from the lesions *Microsporum albicans* and *Microsporum lanosum* may occasionally be present

There is an acute inflammatory type of the disease due to *Trichophyton gypseum* and a chronic type due to *Trichophyton purpureum*

The inflammatory type (*Trichophyton gypseum*) commences on the feet and is manifested by vesicles between the toes or on the soles The skin between the fourth and the fifth toes is very commonly affected The vesicles or bullae rupture and the skin at the site of the lesion becomes macerated soggy and white This appearance may be *maintained for weeks* months or years On peeling away of the soggy white debris of skin fissuring and cracking and erythema occur

During the hot weather there may occur a pruritus of the toes some swelling and the appearance of vesicles on the soles and perhaps on the sides of the toes and

feet Synchronously with the evidence of local inflammation on the feet vesicles may appear on the palms of the hands and sides of the fingers These eruptions are due to the dissemination of products of fungi through the blood stream and the lesions are known as dermatophytids They are sterile on culture

In instances in which there is an increased sensitivity the eruption may spread to involve the intertriginous areas and folds of the upper parts of the thighs the perianal region the axillae the inframammary regions and the umbilicus A secondary infection of the involved areas by pyogenic organisms is common This secondary infection may overshadow the characteristics of the fungous disease and appear as an eczematoid dermatitis

Infection of a nail with *Trichophyton gypseum* may be superficial causing only a white patch on the surface or in the substance of the affected nail (leukonychia trichophytica) A more severe infection causes the nail to turn pale yellow opaque lusterless and friable and separation of the nail may occur Subungual hyperkeratosis *and uneven dystrophic changes in the nail are frequent but paronychia is rare* Mild to severe pain may occur

The appearance of the eruption (with itching) on the feet and on the hands is peculiar to *Trichophyton purpureum* infections Lesions of the feet and hands may occur on the soles and palms the sides the dorsa the toes and fingers or the nails

Of patients who have *Trichophyton gypseum* infections about 7 of every 10 give positive reactions to skin tests with trichophytin in 48 hours of those who have *Trichophyton purpureum* infections about 3 of each 10 give positive reactions after 48 hours Reactions to the trichophytin skin test in those who have dermaphytosis from other organisms are so irregularly present that the value of the test for pur poses of diagnosis is doubtful

The diagnosis is made by culture and identification of the offending organism In most cases of inflammatory tinea the reaction to the trichophytin test is positive When *Trichophyton gypseum* is the causative fungus the prognosis is hopeful If *Trichophyton purpureum* is the offending microorganism cure is difficult The duration of the infection and the extent of involvement of the skin are often deter minative factors in the prognosis

Tinea Versicolor (*Pityriasis Versicolor Chromophytosis*) The microorganism a species of fungus that causes tinea versicolor is known as *Malassezia furfur* The disease affects adults of both sexes The lack of personal hygiene sometimes seems to be a predisposing cause in many instances of the disease

The disease manifests itself by light yellow colored scaly macules and patches starting from barely visible lesions in single or multiple foci The sites of predilection are the thorax the abdomen and the back At times the eruption favors inter triginous locations but it may involve large stretches of skin

During the summer or autumn a patient who has tinea versicolor may exhibit apparently depigmented areas on the surfaces of the affected skin which are exposed to sunlight The patches occupy the sites of lesions of tinea versicolor Their color is not the pale white with demarcated edges and with increase of pigment at the periphery as is present in vitiligo The condition appears year after year and despite becoming less noticeable during the winter it will *reappear* during the summer

The lesions of tinea versicolor under filtered ultraviolet rays reveal a fluo rescence Examination for fungi combined with observation of the patient under filtered ultraviolet radiation is diagnostic The ordinary form of tinea versicolor is not confused with any other disease

Erythrasma Erythrasma is a superficial mycosis resembling tinea versicolor The causative fungus is *Actinomyces minutissimus* (*Microsporum minutissimum*) The means of transmission of the infection is unknown The disease is less common

than tinea versicolor Erythrasma is commoner in young men whereas tinea is commoner in old men and the lesions of erythrasma are limited to one or more in number situated in the axillae the groins the intergluteal cleft or other intertriginous areas The lesion commences as a small scaly, well circumscribed macule with a reddened border which gradually enlarges to form variously sized patches The color varies through mixtures of red and brown, the exact shade depending on the amount of pigment in the skin the age of the lesion (the older the darker) and the amount of sunlight to which the lesion has been subjected

In all cases a mycologic diagnosis should be made by identification of the organism The demonstration of the microorganism may be difficult

Otomycosis (*Myringomycosis*) Otomycosis affects the external ear and the aural canal This condition may be due either to fungi or to streptococci No age is exempt although the majority of patients are young adults

The external ear around the meatus is swollen and red A moist mass of debris fills the canal When this is removed the affected skin is seen to be exudative The disorder may extend down the canal and affect the drum

Lepothrix (*Trichomycosis Axillaris*) Trichomycosis axillaris (*Actinomyces tenuis*) is characterized by nodes on the axillary hair often with the formation of red or black pigment Micrococci are said by some to be responsible for the formation of this pigment and not *Actinomyces tenuis*

Irregular concretions form in the axillae and attach themselves to the hair The attachment is firm and the nodes (concretions) are difficult to dislodge The entire circumference of the hair is involved The concretions are usually yellowish red and black varieties are uncommon The concretions exhibit fluorescence under filtered ultraviolet rays

Tinea Nodosa (*Piedra*) This disease caused by *Trichosporon giganteum* is manifested by small hard nodes along the involved hair shaft The disease is confined to scalp hair of women in South America The European and Asiatic varieties affect only the male beard and mustache never the scalp The nodules of both varieties are stony hard and light or dark brown From 1 to 25 nodes may be present on one hair Shaving is a certain cure

Chromoblastomycosis (*Dermatitis Verrucosa*) Three fungi *Hormodendrum pedrosoi Hormodendrum compactum* and *Phialophora verrucosa* are recognized as causing verrucous nodules The tissue reaction consists of granulomatous changes similar to those resulting from the presence of a foreign body a tuberculoid reaction and miliary abscess formation

The lesions are situated on the leg Each lesion commences as a verrucous nodule or an ulcer The nodule may be reddish purplish or brownish After many months or years verrucous masses are formed by coalescence of two or more lesions and their subsequent growth Growth continues until large pedunculated fimbriated cauliflower like masses are formed Infection by bacteria causes a foul discharge and edema of the food and ankle The lesions resemble psoriasis lupus erythematosus and tuberculosis

Mycologic study and the results of inoculation of guinea pigs may reveal distinguishing features of this disease

Moniliasis The causative microorganism of moniliasis is *Candida albicans* The organism is etiologically weak It does not occur on the normal skin It is a common inhabitant of the orifices of the body and the gastrointestinal tract Once *Candida albicans* is established it may cause either superficial or deep mycotic infections Only the skin infections will be considered here

Housewives bartenders waiters and bakers appear to be more susceptible than other persons to the infection because of their occupational sweating and often wetted hands When moniliasis (*Candida albicans*) infections are limited to a localized area of skin they are termed the moniliid type

These localized affections of the *skin* are manifested by well defined bright red exuding patches with scalloped borders occurring mainly in intertriginous areas Bordering the zone of intertrigo small flaccid vesicopustules may be present A monilial intertrigo occurs in the axillae the groins and the intergluteal fold The process may extend from a primarily intertriginous location to the flat skin and large surfaces of skin of a susceptible person may be affected

Candida albicans infections at the angles of the mouth may occur as rhagades or perleche the bases of which are bright red This type of perleche cannot be easily distinguished from those types due to vitamin deficiency secondarily infected by streptococci

Onychia and paronychia of the *fingernails* and occasionally of the *toenails* occur Infection of multiple digits is the rule Rarely is only one digit involved As a rule the paronychial tissues are the first to be affected A thin purulent discharge may appear under the nail fold Transverse ridges appear on the nail though it remains hard The nail gradually becomes thickened and distorted at the edges and may become yellow The natural glazed surface is unaffected The edges of the nail may be eroded There is no pain except by pressure on the nail and from the paronychia when dry

Thrush is a manifestation of *Candida albicans* It is most commonly seen in infants It may have been contracted at the time of birth from a *Candida albicans* vaginitis of the mother A whitish loosely adherent membrane is attached to the inner surface of the cheeks or to the palate and sometimes to other portions of the oral mucosa The whitish membrane resembles the curd of milk and is distinguish able by the fact that it can be wiped away without leaving residual surface markings

The presence of *Candida albicans* in the *vagina* usually does not indicate the presence of a *Candida albicans* vaginitis In women sufficiently susceptible the organism may produce pruritus and vaginitis accompanied with a thin vaginal dis charge It is even less likely that *Candida albicans* is of etiologic importance in *pruritus ani*

Monilid lesions may occur as sterile vesicular lesions on the *hands* and localized or widespread erythematous vesicular exudative patches caused by dissemination through the blood stream of products of *Candida albicans*

The diagnosis is difficult for culturing of *Candida albicans* from a lesion does not identify the lesion as being caused by *Candida albicans* The disease no doubt occurs but to make the diagnosis is a dangerous practice except when the lesions are around the nails of the hands Identical lesions about the nails may be caused by bacteria The oidiomycin test is of no value in differential diagnosis When infection is gener alized the outcome is often fatal (see Deep Mycotic Infections Chapter 17)

CYSTS SINUSES AND FISTULAS

Cysts sinuses and fistulas are of common association with the cutaneous struc tures The commonest cysts are the epidermal cysts

Anderson classified cysts sinuses and fistulas as they are here described

Epidermal Cysts Epidermal cysts have an epidermal lining but lack definite dermal structures Epidermal cysts are generally found in the cutis or corium and are only rarely situated below the cutis in the subcutaneous tissue Occasionally such cysts may be attached to the epidermis by a band of epithelial composition The great majority however are not connected with either the overlying epidermis or any cutaneous gland or structure

Degenerative changes occur with considerable frequency in epidermal cysts Rupture of the epithelial capsule is common and a foreign body giant cell inflammatory reaction ensues Fibrosis and relative involution may occur and while calci fication is frequent true ossification is rare Malignant degeneration may occur (epithelioma)

Sebaceous Cysts Sebaceous cysts result from the occlusion of sebaceous glands and hence their epithelial lining is composed of secreting sebaceous gland cells The contents of the true sebaceous cyst are light yellow in color homogeneous in consistency and have a disagreeable distinctive odor

The multiple cysts occurring on the trunk in association with acne and seborrhea are true sebaceous cysts

Sweat Duct Cysts Sweat duct cysts are associated with the sweat ducts and sweat glands and are generally termed hydrocystomas or sudoriparous cysts These cysts appear on the cheeks as solitary shiny translucent and fairly deep lesions 1 mm in diameter Occasionally larger ones occur on the upper eyelid The larger solitary lesions are best removed surgically the smaller lesions can be destroyed by electrodesiccation

Dermoid Cysts True dermoid cysts are congenital in origin and are situated in areas where there was a connection with embryologic fissures and clefts They occur in the cutis or the corium They may occur at either the inner or the outer canthus of the eye in relation to the bony walls of the orbit or on the midline of the nose along the lines of closure of the nasal bones or along the anterior midline of the neck

The history of a *dermoid cyst of the neck* is that of a midline swelling in the neck above the thyroid gland which has been present for years Either because of an increase in size or for cosmetic reasons the patient consults a physician The diagnosis of a cyst or a mole is made and there is an attempt to treat the lesion either by electrodesiccation or by surgical removal A few hairs are generally removed at such times There results a sinus opening which occasionally discharges cheesy material pus and blood The conversion of the dermoid to a sinus opening therefore results from subtotal surgical removal or at times may take place spontaneously

These midline dermoids of the neck may grossly resemble thyroglossal duct cysts Thyroglossal duct cysts are remnants of the embryonic duct which connected the thyroid gland to the tongue The remnant of the lingual connection in adults is the foramen caecum in its posterior dorsal surface Thyroglossal duct cysts may appear as cystic masses on the posterior dorsal surface of the tongue

Mucous Cysts The so called mucous retention cysts which are situated on the inner aspect of the lower lip usually opposite the cuspid tooth are very common (see Chapter 4)

Traumatic Epithelial Cysts Cysts resulting from trauma are caused by (1) displacement of epithelial islands (2) stimulation of epithelial growth or (3) production of alterations in the normal structure of cutaneous appendages such as the hair sebaceous and sweat glands

A traumatic cyst occurring under the free end of the fingernail may cause moderate tenderness and even erode the terminal phalanx by pressure Ossification may occur within the cyst and necessitate removal

Sinuses and Fistulas A sinus is an open channel tract or tube blind at one end and communicating at the other with a cutaneous or mucous surface A fistula may be defined as an open channel between two body cavities or between a cavity and the skin or between skin and skin It is lined with either granulation tissue or epithelium and limited by an inflammatory membrane A sinus tract is for example what generally occurs with dermoid cysts

Congenital Sinuses of the Lips Unilateral or bilateral millimeter sized slitlike openings of congenital origin occur on the lower lip If bilateral there is one opening on either side of the midline The sinus tracts are generally 1 to 2 cm long and extend downward and inward toward the midline generally ending in a blind pocket immediately under the mucous membrane of the inner surface of the lip

These bilateral sinuses of the lip are called fistulas and though rare tend to be

familial and are often accompanied by other deformities such as cleft palate and harelip

The solitary midline sinus occurring on the upper lip just beneath the columella of the nose is the external manifestation of a deeper lying dermoid cyst

Epidermal Fistulas In their simplest form epidermal fistulas are seen about the face neck buttocks and inner aspects of the thighs and less commonly about the breasts and axillae The process either originates in the double comedones of a severe acne vulgaris or follows a process similar to hidradenitis suppurativa

Congenital auricular fistula sinus preauricularis and fistula auris congenita occur as small papules situated at the anterior border of the ascending limb of the helix From such sinus openings there may be an occasional offensive discharge At other times with the closure of the orifice a retention cyst may develop in front of the ear Finally secondary infection may convert the sinus tract or cyst into a walled abscess which opens either spontaneously or by incision In either event a solitary *granulomatous lesion anterior to the ear ensues As the result of repeated infections* many fistulous tracts develop in an extensive area of skin This fistulous area may present a lesion which resembles lupus vulgaris or actinomycosis

There is a frequent association of preauricular sinus a developmental defect with other congenital malformations such as branchial fistulas of the neck hypospadias and congenital absence of the tear duct

Sinus tracts of dental origin are common and are often associated with an alveolar abscess at the root of a tooth less commonly with syphilis of the mandible and rarely with adamantinoma of the jaw They may open on the chin the cheek the floor of the nose near the orbit and on the buccal or lingual aspects of the gums or at distant sites as the neck

On the lateral aspects of the neck sides of the face and as high as the lower part of the eye may occur cysts which are remnants of an incomplete closure of an embryonic branchial cleft

Diagnosis of Cysts Sinuses and Fistulas Aside from the clinical findings visualization of the tract and cysts by means of roentgenograms after the introduction of radiopaque solutions is a helpful measure In some cases much evidence as to the place of opening of a fistula may be obtained by the injection of a solution of methylene blue

Surgical treatment of some of these lesions may be difficult A final diagnosis awaits histologic studies

INFECTIONS OF THE SKIN BY HIGHER ANIMAL PARASITES

Helminths The term helminths is used to designate wormlike animals of three diverse phyla (1) Annelida (segmented worms) (2) Nemathelminthes (round worms) and (3) Platyhelminthes (flatworms) In medical parasitology all parasitic worms are collectively referred to as helminths

Annelida The segmented worms are mostly free living The ectoparasitic *leeches* comprise the parasitic members of this phylum

The chief predators of man and animals are the terrestrial leeches that live in damp tropical forests where they attach themselves to anyone within reach Various species have been described in southern Asia the East Indies Australia Oceania and South America Aquatic species attack bathers examples of these leeches are those that are encountered in the swimming holes along some southern streams and elsewhere in the United States

The bite usually unnoticed is detected by the presence of blood or the engorged leech The wound though painless bleeds readily because of the anticoagulative secretions of the leech and heals slowly Heavy infestations of man and animals may result in serious exsanguination A number of cases of infestation of

the upper part of the respiratory tract and of the digestive tract from drinking water have been reported Leeches in these locations after narcotization may be removed with foreceps Those attached to the exterior of the body may be removed after application of vinegar or strong salt solution to loosen their hold

Creeping Eruption (*Hookworm Disease Ancylostomiasis*) In man the skin lesions of creeping eruption are caused by penetrations of the larvae commonly known as hookworms of at least three species of nematode worms of the family Ancylostomatidae (superfamily Strongyloidea) *Ancylostoma duodenale Necator americanus* and *Ancylostoma braziliense*

The eruption develops after contact of the patient with moist sand and earth contaminated by the feces of infected dogs cats or human beings At the point of invasion a reddish itchy papule develops and within two or three days narrow linear slightly elevated erythematous serpiginous intracutaneous tunnels are produced by the migration of the larvae which move from a fraction of an inch to several inches each day Vesicles form along their course and the surface becomes dry and crusty The itching is intense and the resultant scratching may lead to secondary infection The commonest site is on the hands or feet but the eruption may occur on any part of the body It persists for several days or weeks

Dracunculiasis The Dracunculidae are long slender nematode worms filarial parasites The definitive hosts other than man in North America are the fox raccoon and mink The species parasitic in man is *Dracunculus medinensis* of the family Dracunculidae

When the infected cyclops is ingested in drinking water the larvae penetrate the wall of the digestive tract and migrate to the loose connective tissues When the female worm reaches the surface of the body it liberates a toxic substance into the subdermal tissues producing a local inflammatory reaction in the form of a subacute sterile abscess with serous exudation and formation of blisters

The early manifestations of urticaria erythema dyspnea vomiting diarrhea pruritus and giddiness are caused by the absorption of allergenic substances from the worm If the worm is broken during extraction and the larvae escape into the subcutaneous tissues a severe inflammatory reaction with fever ensues and if secondary bacterial infection occurs formation of abscess sloughing of the tissues and even fatal septicemia may result

The local cutaneous lesion appears at first as a reddish papule with a vesicular center and indurated margin The blister contains a clear yellow fluid The worm lies in a subcutaneous tunnel with its anterior end beneath the blister Its course may be marked by induration and edema Contamination of the ruptured blister by pyogenic bacteria may produce abscesses cellulitis and ulcers At times the worm may become calcified and remain for years as a hard twisted cord beneath the skin

Diagnosis is made from the local lesion worm and larvae Dead or calcified worms may be located by roentgenologic examination Treatment involves the extirpation or destruction of the worm The prognosis is usually good

Cercarial Dermatitis The cercariae which are larvae of certain species of animal schistosomes or *flukes (Platyhelminthes) produce in man a* cercarial dermatitis

The cercariae of 6 species of animal schistosomes have been associated with cercarial dermatitis (1) *Cercaria elvae* (2) *Cercaria stagnicolae* (3) *Cercaria physellae* (4) *Schistosoma deuthitti* (5) *Trichobilharzia ocellata* and (6) *Schistosoma spindale* The first four are found in the United States of America

In the United States *Cercaria elvae* and *Cercaria physellae* found in swampy waters produce *collectors itch Cercaria stagnicolae* which frequents the shores of lakes is more often the cause of *swimmers itch* Persons in contact with aquatic vegetation are most likely to be infected with *Cercaria elvae* and *Cercaria physellae*

The affected beaches are usually near vegetation that furnishes a favorable habitat for snails The cercariae swarm in the shallow water or are swept toward the shore by wave action They survive about 24 hours

Cercarial dermatitis has been recorded in England France Germany Switzerland the Federated Malay States Canada and the United States of America In North America endemic centers have been found in Saskatchewan and Manitoba and in Wisconsin Michigan Minnesota and Iowa Outbreaks have occurred in Illinois North Dakota Nebraska Texas Florida and Washington

The cercariae adhere with the ventral suckers and enter in about 5 minutes through the action of their anterior spines and lytic secretions Penetration of the skin occurs when the film of water evaporates The cercariae may remain alive in place for a day but the reaction persists around the burrows They may be destroyed in the epithelial layers of the skin and become walled off During these actions there is inflammatory response with edema

As the water evaporates a prickling sensation is followed by the rapid development of urticarial wheals which subside in about half an hour leaving a few minute macules After some hours severe itching edema and the transformation of the macules into papules and occasional pustules occur reaching maximal intensity in two or three days The papular and sometimes hemorrhagic rash heals in a week or more but may be complicated by scratching and secondary infection Individuals vary in susceptibility

THE DERMATOSES

• The term dermatosis is all inclusive It simply means a skin disease The term toxic dermatosis is all inclusive too for it refers to any skin disorder referable to a toxic agent Still more tolerance is required in the use of these terms for there are (1) the dermatoses due to a known or a proved group of toxic agents and (2) the dermatoses due to unknown toxic agents

The known agents which may produce dermatosis are (1) bacteria (2) some higher plant and animal parasites (3) viruses (4) chemical agents and (5) physical agents These agents have different methods of attack Some are able to attack directly while others attack indirectly The latter group make themselves manifest through sensitization acting as allergens on an individual predestined by inheriting the weakness of being easily sensitized (see Chapter 18)

The various dermatoses are mentioned in their relation to a disease or in association with a toxic agent or agents in various parts of this text

ENDOCRINE DISTURBANCES OF THE SKIN

Seborrhea A definite etiology has not been established for seborrhea

At puberty the adult distribution and type of hair develop and there is an increase and perhaps a change in the quality of the sebaceous secretion These changes are synchronous with puberty and are thus attributed to functioning of the glands of internal secretion It is in early adulthood that dandruff begins to be troublesome in some individuals

Seborrhea is often described as being of two types The dry type or so called *seborrhea sicca* is commonly known as *dandruff* The oily type is the so called *seborrhea oleosa* It frequently is impossible to differentiate the two types Seborrhea affects the scalp the eyebrows the central portion of the face and the central portion of the thorax the anterior and posterior axillae and the bends of the elbows In women it sometimes may be found about the clitoris The disease may be associated with acne vulgaris The eruption appears as dull reddish erythematous patches often with greasy scales often well defined on the body and it sometimes forms annular lesions over the sternum Itching may not be very annoying except just before retiring and during the night The patient may awake to find himself scratch

ing the scalp Occasionally the dermatitis extends down from the scalp or from the hairline to the forehead The patch may be well defined at its borders

Acne Vulgaris The disease is manifested as an inflammatory affection of the sebaceous glands The characteristic lesions the comedo the papule and the pustule are distributed over the face trunk and buttocks where the sebaceous glands are largest The lesions are painless unless severely infected In boys the lesions begin often on the nose and chin synchronously with puberty and in girls with the onset of menstruation

The primary infection has been attributed to *Bacillus acnes* (*Corynebacterium acnes*) It is secondary invasion of the lesions by staphylococci that leads to pustulation The altered sebaceous secretion in the comedones turns black on exposure to air When a comedo is expressed a small plug or wormlike mass of material exudes or shoots from the follicle to be followed by thin pus Around the papule and the pustule is well marked hyperemia Sometimes the pustules increase greatly in size and furuncles develop Chronicity results in deep scars which on the face may be as disfiguring as those of severe smallpox If occlusion of the follicular openings takes place sebaceous cysts develop

Acne is differentiated from the drug eruptions by the history Iodides bromides and various estrogenic preparations or hormones taken internally and tar paraffin and chlorine when applied externally may produce lesions almost identical to acne although not always on the usual sites

After the patient has reached the age of 30 years acne vulgaris tends to heal gradually and the scarring is less conspicuous by the time the age of 40 years is reached

Rosacea The occurrence of rosacea about the time of the menopause during the menopausal flushing of the face is etiologically suggestive of an endocrine association

Rosacea differs from acne vulgaris in the absence of comedones in distribution and in age of incidence Often rosacea begins in the late twenties or the onset may be delayed until the forties It commences as a chronic hyperemia of the face which often terminates in a permanent dilatation of the capillaries During the course of this hyperemia papules pustules and a patchy scaliness occur The distribution of the disease is on the nose cheeks chin and forehead especially between the eyes

Rhinophyma is the advanced or the extreme stage of rosacea Its worst manifestations are seen in alcoholics Often the rosacea is accompanied by an enlargement of the tip of the nose which becomes irregularly bulbous forming a soft fleshy mass pitted with dilated follicles from which sebaceous material can be expressed The organism *Demodex folliculorum* found in these dilated follicles has no etiologic significance

Senile Dermatoses As age advances the skin loses its elasticity becomes lax wrinkled lusterless and seems too big Warts that resemble flat warts but usually are larger and have a dark surface often appear over the neck and the trunk These growths do not become malignant

Senile keratoses little hard persistent areas of scaliness on the face nose and hands are evidence of the effects on the skin from years of exposure to sun and wind The skin of those who have red hair is more susceptible and more sensitive to these influences than skin of other coloration The extreme of sensitivity is shown in xeroderma pigmentosum and similar conditions may be produced by roentgen rays or radium in a much shorter time They are of the same nature however and all have a tendency to become malignant

Hypothyroidism Deficiency in thyroid secretion leads to decrease in the secretions of the skin glands and produces a dry skin with increase in the thickness of the horny layer (xeroderma)

These skin changes are improved by thyroid therapy Hypothyroidism may on

occasion be accompanied by an ichthyosis The ichthyosis too is improved by thyroid therapy However a deficient thyroid plays no part in the etiology of ichthyosis

Skin Diabetes Urbach has defined skin diabetes as the syndrome of therapy resistant recurrent or chronic dermatosis a high fasting skin sugar level together with a normal blood sugar curve and pronounced improvement of the skin disease as well as a drop in the high skin sugar level on a low carbohydrate diet sometimes combined with insulin

In support of this view that there is a connection between the clinical syndrome and a disturbance in the carbohydrate metabolism as in diabetes are (1) the high fasting skin sugar levels which attain and even exceed 80 mg per 100 gm (2) the pathologic and characteristically diabetic course of the skin sugar curve following oral sugar feeding (3) the return to normal of the fasting skin sugar level as well as of the skin sugar tolerance curve after a low carbohydrate diet notably in combination with insulin

On the basis of clinical experience a diabetic diet should be tried in cases of therapy resistant skin diseases such as furunculosis eczema or pruritus even when the blood sugar tolerance curve is normal

THE EFFECTS OF DISTURBANCES OF NERVE FUNCTION OR PSYCHIC CONTROL

Pruritus Pruritus is an integral part of the symptom complex of a great many skin disorders The cause of the pruritus may be an obvious organic disease or it may be entirely of functional origin

Pruritus Ani vel Vulvae Pruritus ani and pruritus vulvae are in the large majority of cases purely of functional origin Rarely will diabetes or threadworms (*Enterobius vermicularis*) be found as causative factors Occasionally a local condition such as a vaginal discharge hemorrhoids anal fissure or an intertrigo will be found

Neurodermatitis A neurodermatitis occurs as a well defined patch of thickened lichenified skin often scaly and intensely itchy which has been present for a very long time These lesions occur where they are easily accessible to the patient and therefore can be frequently rubbed or where the clothing rubs Irritation leads to itch and scratching which increases the itch and results finally in the thickened crisscrossed patch

Senile Pruritus This general itchiness of the dry skin of elderly people may be a most distressing condition and may gravely alter their physical and mental well being

SKIN DISEASES OF UNKNOWN ORIGIN

Psoriasis Psoriasis is a common not serious dermatosis of white peoples The lesions occur more frequently and more profusely on parts of the body that are covered with clothing than on parts that are bare They do not cause the patient discomfort

Men are more often affected by the disease than are women and morbid heredity seems to be of etiologic importance Some families are more prone to the disease than are others Lesions of psoriasis may occur along the line of a cut or scratch in a scar or on areas such as the waist where clothing presses Apparently any damage even trivial to the skin may evoke the eruption locally

Psoriasis often becomes manifest during childhood or early adult life but it may commence for the first time at any age from infancy on The disease may be limited for years to a patch on the elbow or knee and then suddenly it appears on other parts of the body The disease may be active while the patient is well and disappear during an illness such as pneumonia or during a pregnancy only to return when the patient regains his normal state of health In some instances an acute exacerba

tion may be activated by a general nervous debility Remissions occur the patient remaining free from the disease for years

Sunlight is beneficial to most of these patients The lesions may largely heal after repeated judicious exposure to sunlight The natural dislike for exposing the affected ugly skin to view however prevents many of the patients from obtaining an adequate amount of sunlight

The lesion of psoriasis is a reddish slightly thickened area of skin covered to a considerable depth with a glistening silvery scale This character of the scale may not be apparent at first sight but it appears if the lesion is scratched As the scratching is continued the scale gradually comes away in masses or as a fine powder until its lowest layer is reached This layer comes away in one piece as a soft scale and leaves a red shiny area dotted with tiny bleeding points Removing the scale as described is truly the *scratch* test and is almost diagnostic in doubtful cases

The size and shape of the lesions of psoriasis vary Descriptive terms are used for the variations among which are the following If the lesions appear acutely as profuse eruptions of small distinct and irregular patches the condition is *psoriasis guttata* The spots may enlarge into sizable patches as in *psoriasis nummularis* If the scales become heaped up the disease is of the rupial form *psoriasis rup oides* In some cases the center of the lesion clears while the edge remains active and circinate or serpiginous *psoriasis circinata* and *psoriasis gyrata* The size of the patch is roughly indicative of its age and in some long standing cases the patches may cover large areas of the body and limbs They may have a maplike appearance *psoriasis geographica* In all cases however the scratch test indicates the nature of the disease

The lesions usually occur on the extensor surfaces and almost always the elbows and knees and the scalp are affected sites On the scalp the hair prevents the scales from being easily shed so that the disease in this area is more easily discovered by palpation Lesions elsewhere usually will make the diagnosis of scalp lesions clear the resemblance of psoriasis to seborrhea is not great Psoriasis may be found behind the ears and under the breasts

The nails may be affected giving rise to irregular thickening of one or more nails There may be rows of punctate depressions in the nails In cases of psoriatic arthritis the lesions of the nails are sometimes the only ones present

In rare instances exposed parts may be affected and lesions may appear on the face and hands Psoriasis of the palms and soles may assume a pustular form

In some instances psoriasis appears to be associated with rheumatoid arthritis Both psoriasis and rheumatoid arthritis are common diseases and some of the apparent association may be due to chance

In doubtful cases differential diagnosis is made more easily if the scratch test is used repeatedly Syphilitic lesions give a feeling of greater thickening of the skin are more reddish brown than psoriatic lesions and show involvement of the face and the forehead Circinate psoriasis simulates ringworm but the scales do not contain fungus Seborrheic pityriasis of the scalp may offer difficulty the localized heaped up patches of psoriasis palpable to the touch and the scratch test help to differentiate the two conditions Psoriasis of the palms resembles a ringworm infection and so may psoriasis of the nails so that examination of scrapings for fungus is necessary Psoriasis and lichen planus may resemble each other

Prognosis is uncertain The disease may last for a lifetime An acute attack may necessitate the patient s resting from work

The laity does not know that the disease is noncontagious The patients should be discouraged from use of public swimming facilities and from military service

Lichen (Rubor) Planus Lichen (rubor) planus is a rare chronic benign disease occurring in middle aged individuals which may persist for the rest of their lives It can and does appear in an acute form with a generalized eruption which is accompanied by itching It may disappear spontaneously after weeks or months Women are more frequently affected than men The general health remai

Itching is not often severe though it may become severe enough to interfere with sleep Oral lesions may be painful enough to interfere with eating

The lesion is a polygonal flat topped shiny crisscrossed papule which breaks the fine lines of the skin The papule varies in size sometimes reaching a few millimeters in diameter but agglomerations of papules may spread over a large area The light lavender color is characteristic of the lesions and may be observed at the edges of chronic patches if the scales are wetted away A feature of the disease is the occurrence of the lesions along the lines of the ribs or places of skin trauma for instance where a belt or a corset or other clothing rubs the skin

The general appearance of the lesion varies considerably The acute lesions consist almost entirely of papules Variations in secondary characteristics have occasioned descriptions of the following forms of the disease

The *bullous* variety is an acute form characterized by bullae interspersed with the papules The *annular* variety is characterized by rings of papules caused by the regression of the central lesions and the appearance of fresh papules in the periphery The *hypertrophic* or *warty* variety is the characteristic manifestation of chronic lichen planus This form may resemble psoriasis if the characteristic papules and violaceous color at the edges are not evident The *atrophic* variety is rare The patches are ill developed and the macular lesions are purplish or white frequently striated and sometimes surrounded by erythema

The mucous membranes of the mouth or the vagina may be affected giving rise to leukoplakia like lesions especially if the tongue is involved These lesions of mucous membrane often cause considerable discomfort

The flexor aspects of the forearms and the fronts of the legs and around the knees and over the sacrum are favorite sites of eruption However the eruption may be widespread especially in acute attacks

The disease may run a short course of a few weeks or months and the patient gets well It is the chronic form with chronic warty patches giving rise to discomfort by their presence on the legs in the mouth or vagina or on the glans penis which most frequently is seen by the physician

The essential lesions of lichen planus and psoriasis are different and the distribution varies in the two diseases In lichen planus the papules are on the front of the forearms and often there are patches in the mouth In psoriasis the knees elbows and scalp are involved Chronic scaly patches below the knees are found in both diseases and if no lesions are present elsewhere the diagnosis depends on local characteristics Psoriatic patches have little or no itch and have a shiny scaliness which gives a positive reaction to the scratch test (p 512) Chronic patches of lichen planus are itchy in some cases intensely so *the scale is not easily* removed and the violet color is characteristic

A patch of neurodermatitis may simulate chronic lichen planus A neurodermite may be very itchy purplish or brown with adherent scale and cross hatched with lines but its distribution is somewhat different from that of lichen planus The neurodermite is usually where it can be reached easily The papules typical of lichen planus are not present in the lesions of neurodermatitis or elsewhere on the body and the violaceous halo of the lichen planus patch is absent

The lesions of mucous membrane must be distinguished from leukoplakia The lesions of the mucous membrane especially of the penis or of the skin may resemble syphilis

Pityriasis Rosea This disease resembles a dermatitis due to an infection It is most prevalent in young adults The herald patch which has been considered the primary lesion can be identified in about 3 of 4 cases It often occurs on the lower part of the abdomen or on the flanks The herald patch *appears before* the rest of the eruption and may pass unnoticed since it is usually symptomless It is distinguished by its greater size

The essential lesion including the herald patch is an oval rose pink macule

which enlarges and sheds fine scales as the center clears leaving scales at the periphery The long axis of the patches on the body is generally parallel to the line of the ribs The lesions may vary in average size and a papular form may occur Vesiculation is a rare occurrence

The herald patch is followed in 2 or 3 days sometimes longer by the general eruption which often is confined to the trunk and the upper parts of the limbs General symptoms are seldom evident though an initial low fever may be present Usually the eruption fades and in from 4 to 6 weeks from the onset has disappeared although it may persist for months The eruption is not itchy One attack is thought to confer immunity

The ringed scaly lesions may suggest ringworm but fungus is not present Seborrheic pityriasis and syphilis do not offer much difficulty in differentiation

Pityriasis Rubra Pilaris This rare condition is characterized by the appearance of reddish papules corresponding to the hair follicles over the limbs and face and the middle phalanges of the fingers Elsewhere the eruption may closely resemble psoriasis From psoriasis it is differentiated by the scratch test

Exfoliative Dermatitis (Pityriasis) An exfoliative dermatitis is characterized by redness and profuse scaling of the skin It may occur secondary to many skin conditions such as psoriasis pityriasis rubra pilaris drug intolerances and poisoning by heavy metals such as organic arsenical compounds In a great many instances it is a reaction to overtreatment of a skin disorder The patient often is obviously ill The disease runs a chronic course

The exfoliation is first manifest on intertriginous surfaces as the axillae or groins and spreads quickly so that the entire integument may become involved The skin is thickened and red and there is profuse desquamation of large lamellar scales The degree of severity of the eruption varies considerably The desquamation becomes less In severe instances the eruption lasts for months and especially in the elderly the disease frequently is fatal

Indeterminate Vesicular Lesions of the Hands *Cheiropompholyx* Cheiropompholyx appears as minute vesicles often discrete on the hands along the sides of the fingers and in the palms of the hands and on the soles of the feet The vesicles come to the surface of the palms and fingers burst and leave a raw weeping surface which heals slowly and thus may be subjected to much irritation which impedes healing The slow healing requires that the condition be distinguished from ringworm infection A differentiation from an infective dermatitis (infectious eczematoid dermatitis) cannot usually be made

NEW GROWTHS

Benign Growths *Callosities* Callosities are thickenings of epithelium due to friction heat pressure irritation and perspiration They may occur on any part of the body which is subjected to the foregoing forces The hands are favorite sites for instance the horny hand of toil but the callosities occur on the knees of the devout on the buttocks of the lazy and on the feet of those who walk a great deal

Callus and Corns (*Clavus*) As a rule corns are the result of pressure and friction from ill fitting shoes which crowd the toes together and force them into a hammer toe position Hammer toe itself may be free of corns Faulty foot posture with concentration of weight stresses on certain areas of the foot is another causative factor Small exostoses at the joint margins of the interphalangeal joints cause clavus formation from pressure on an adjoining toe

As the horny layers of the corn become thickened the apex is forced deeper into the sensitive papillae of the corium and discomfort and pain result Objectively there is a circumscribed callus which when trimmed away reveals a horny mass extending as a cone downward into the deeper areas

Soft Corns Soft corns occur on the lateral surfaces of and in the web between the fourth and fifth toes and the bony prominence is nearly always on the base of the first phalanx of the fourth toe They do not become hard and dry but are soft and elastic because of moisture and maceration for this reason they are called soft corns

Verruca Plantaris Plantar warts are infectious in origin and once infection has occurred they tend to spread to other parts of the foot Plantar warts apparently seldom occur except at the site of a callus

Plantar warts are small circumscribed callus like growths which form on the plantar surface of the foot and about the heel They are extraordinarily sensitive to pressure and thus may produce discomfort seemingly out of all proportion to their size Plantar warts differ materially from callosities and corns Shave off the top of the horny callous layer of a plantar wart and a central core composed of hyper trophied papillae which is soft and vascular and has a marked tendency to bleed will be found

Fibromas In the majority of cases fibromas arise from sebaceous glands The most marked type is seen in the molluscum fibrosum pendulosum or fibroma molluscum (Recklinghausen s disease) which when extensive may cover most of the trunk with large pendulous growths sometimes cystic at other times like emptied sacks at others like small cystic swellings These are often called neuro fibromas which they are not

Osteomas Osteomas of the skin are often secondary ossifications and often originate from injuries inflammatory processes or tumors In rare instances osteomas of the skin occur as congenital abnormalities

Keloid Keloid is an overgrowth of scarred tissue It may have resulted from (1) an intense inflammatory reaction or (2) innate susceptibility peculiar to certain skins It is more nearly related to the texture of the skin than to the color Keloids are likely to follow burns These growths often occur in operative scars even small scars and tend to extend beyond the limits of the original scars In the beginning there may be pain and tenderness on pressure In time a year or two a keloid tends to disappear

Xanthelasma (*Xanthoma*) Xanthelasma is a form of xanthoma characterized by soft light leather-colored or yellowish patches appearing especially on the eyelids near the inner canthi (see Diseases of Cellular Lipid Metabolism in Chapter 22) These yellow nodules in the skin have no relation to diabetes (xanthoma dia beticorum)

Keratosis Senilis Over the face neck back and hands of aging patients appear flat moderately adherent slightly raised yellowish brown to dark gray or almost black lesions When forcibly removed they show horny prolongations on their under surfaces Some families who have freckles are more prone to have these lesions than are others Keratosis senilis has little or no tendency to undergo malignant changes

Malignant Growths ***Epithelioma*** Epitheliomas of the skin comprise (1) the basal cell type (rodent ulcer) and (2) the prickle cell type (squamous epithelioma) In addition the pigmented epithelioma (melano-epithelioma and nevocarcinoma) may occur and these are most highly malignant tumors

RODENT ULCER The rodent ulcer basal cell carcinoma is the commonest malignant tumor of the skin These lesions usually multiple (1) arise at the junction of skin and mucous membrane (2) originate as cystic growths benign at first and (3) arise in elderly persons beginning as senile hyperkeratosis The sites of occur rence are usually near the eyelids nose and about the ears but the tumors may occur on any part of the face or hands Generally there is a history of a lesion which commenced as a small papule and enlarged slowly over a period of many years growing much more rapidly after ulceration set in In cases of long standing ulcerat

ing lesions about the face the superior maxilla and the nose may become deeply eroded In these regions the ulceration may if not checked often penetrate into the nasopharynx or up into the eyeball causing great destruction in these regions

Although rodent ulcer may affect children the growth usually commences when the patient is about 45 years of age or older When fully developed the lesion is a small ulcer with an irregular base and a rounded edge with white firm nodules occurring along the margins of the ulcer Superficial lesions may partly heal and scar over No discomfort is present until the deeper tissues are affected when there are severe pains

In some instances this skin lesion will have the features suggestive of syphilis lupus and squamous epithelioma The history of syphilis is shorter than that of rodent ulcer and as the disease generally is tertiary when ulceration is present a search of the skin is made for the presence of other scars and pigmentation Serologic tests of the blood and spinal fluid may be made as aids to diagnosis Lupus generally commences before puberty and while there may be scarring the clear reddish nodules are commonly evident and do not possess the typical rolled edge observable in rodent ulcer Squamous epithelioma is much more rapid in its growth than rodent ulcer and may be accompanied by secondary enlargement of the adjacent lymph nodes which are not affected in rodent ulcer

SQUAMOUS EPITHELIOMA Epithelioma in the early stage often leads to difficulty in diagnosis It differs from rodent ulcer in being more rapid in its spread in showing no healed areas and in producing in time secondary involvement of the lymph nodes

In early stages differentiation between benign and malignant lesions and in later stages distinction between basal and squamous cell carcinoma as well as the identification of rare tumors can be made by biopsy study Biopsy in suspected skin cancer is simple and virtually harmless (except in the cutaneous surface in mucous membranes and in the eye)

The prognosis depends on the size of the lesion and the adequacy of the treatment Small localized lesions without invasion 1 cm or less in diameter are cured in 9 out of each 10 cases When lesions are 1 to 2 5 cm in diameter without deep infiltration less than one half of five year arrests may be expected Lesions more than 2 5 cm in diameter with evidence of invasion of muscle bone or cartilage or with involvement of regional lymph nodes are rarely curable but significant palliation can usually be obtained through the use of protracted roentgen therapy by means of electrocoagulation or actual cautery or by other appropriate palliative surgical procedures However some five year arrests can be expected

Malignant Melanomas (*Moles and Pigmented Nevi*) Malignant melanomas may develop from pigmented moles of the skin as the result of trauma naturally occurring as from clothing shaving or tight fitting shoes and trauma from incomplete removal A melanoma may become actively malignant during pregnancy However generally a quiescent melanoma or mole simply becomes malignant suddenly and metastasizes to distant parts

When malignant change takes place the mole may increase in size become raised from the skin surface and may become deeply pigmented It becomes inflamed shows a moist surface and bleeding may occur Sometimes a black spot appears in the mole Occasionally a number of pigmented nodules develop around it The lesion becomes ulcerated and as the disease progresses the regional lymph nodes become involved In some instances multiple widespread malignant lesions seem to occur simultaneously

In some instances lesions similar to melanoma may be present These lesions are almost without color If color is present it is a light brown These lesions occur as very small flat nodules or as bulky tumors These lesions are in sharp contrast to the intensely colored ones which are of an intense bluish black to black

There are no reliable criteria for differentiating between benign and early malignant melanomas In general the harmless lesions are more likely to be light in color and soft they may be papillomatous and may be hairy Alteration darkening in color increase in size change in contour or texture arouses the suspicion of malignant activity The appearance of satellite melanotic tumors about the periphery of the original lesion is evidence that dissemination has already occurred

Biopsy of pigmented lesions is definitely contraindicated These lesions should either be widely and deeply excised or left alone It is impracticable and inadvisable to attempt removal of all pigmented nevi from all normal individuals In general lesions subject to frequent trauma situated on the feet genitalia or conjunctiva and in which there is objective evidence of growth inflammation or itching should be excised Once a malignant melanoma has become disseminated only palliative measures are available Melanomas are generally but not always radioresistant

Sarcoma Sarcoma involving the skin but seldom arising from it occasionally is seen in nodular form occurring on various parts of the body These tumors are highly malignant as evidenced by rapid formation reddish color and tendency to ulcerate easily These growths may be recognized early but not soon enough for the prognosis irrespective of treatment is bad

Kaposi s sarcoma occurs as multiple hemorrhagic purpuric appearing lesions generally affecting the lower extremities of persons between the ages of 40 years and 50 years The course is variable but is less hazardous than that of the single sarcoma

Kaposi s sarcoma has to be differentiated from purpura hemorrhagica by a study of the blood for those findings diagnostic of purpura (see the Purpuras in Chapter 12) The diagnosis is made by histologic study of biopsied material

SUBCUTANEOUS TISSUES

Diseases of the subcutaneous tissue are usually considered with those of the skin or of the more deeply situated structures for interposed as these tissues are between the skin and more deeply lying structures they may be affected in disease of either

The Reticuloses The term reticuloses designates those conditions of the skin which may affect the subcutaneous tissues through involvement of the reticulo endothelial cells of the skin and the lymph nodes

The reticuloses include the leukemias the lymphoblastomas (Hodgkin s disease) mycosis fungoides and lipolemic reticulosis In the leukemias and the lymphoblastomas there occasionally are lesions of the skin whereas in mycosis fungoides the skin manifestations are the characteristic feature of the disease

In the reticuloses granulomatous lesions are relatively rare in the skin It appears that in the initial states the tissue reaction in the skin is more acute in type than that in the lymph nodes However fibrosis and the development of Sternberg Reed cells are seldom exhibited in the skin Lymph nodes therefore are far more promising leads toward histologic diagnosis than the skin The eosinophilia in some cases may or may not be significant of an interrelationship with eosinophilic granuloma

The cutaneous lesions when present in lymphatic leukemia and lymphadenoma consist of pruritus urticaria or a nodular purple or reddish brown eruption A general exfoliative dermatitis may occur especially in Hodgkin s disease The diagnosis can be made only from histologic studies of sections of the affected skin

Lipolemic Reticulosis Lipolemic reticulosis may resemble Hodgkin s disease and other reticuloses The histologic changes in the lymph nodes are constant There is disorganization of the normal structure of the nodes though this is usually spotty In those areas the lymph sinuses are dilated and often empty There is proliferation of the reticular tissue and many eosinophils and histiocytes are present Melanin is

present in large amounts both free and in melanophores Fat stains show considerable lipoid material

The usual clinical manifestation is a generalized lichenified or eczematous eruption of several years' duration with darkening of the skin In Negroes there may be depigmentation Often there is loss of hair, especially of the scalp and axillae Frequently there are changes in the nails with thickening and discoloration and subungual thickening and hyperkeratosis

Lipomas Lipomas occur as lobulated or rounded masses in the subcutaneous tissue They are soft and freely movable When traction is made on the overlying skin, multiple small dimples form in it Hard lipomas are designated fibrolipomas whereas those having an increased vascularity may be called angiolipomas

These tumors may occur anywhere in the body but are most commonly situated in the subcutaneous tissues over the back shoulders neck thorax abdomen and buttocks and legs Other sites for lipomas are the mesentery, and serous tissues of the whole alimentary tract kidneys heart uterus meninges respiratory passages and occasionally in the mouth and tongue

Lipomatosis Multiple localized or unusual depositions of fat are classified as lipomatosis to distinguish them from the single localized fatty tumors and form of obesity They cause little concern except from a cosmetic point of view or when they mechanically interfere with normal function because of being inopportunely situated so that function may be disturbed

Heredity is an etiologic factor in the symptomatic progressive forms of lipomatosis Lipomatosis of the pseudohypertrophic form of muscular dystrophy in which the adipose tissue invades between the muscle bundles causes some of the abnormal enlargement of the legs in these children Lipodystrophia progressiva a rare condition of girl children is characterized by complete disappearance of subcutaneous fat above the hips and the development of psychoneurotic symptoms In the nodular circumscribed lipomatosis the lesions are distributed as firm discrete freely movable subcutaneous tumors over the forearms thighs and often the back The diagnosis is usually obvious but a biopsy study may be necessary for confirmation

Adiposis Dolorosa (Dercum s Disease) Dercum s disease or adiposis dolorosa is a syndrome consisting of tender and painful fat nodules often attaining large size situated in the femoral and inguinal regions or in the cervical region When the nodules are situated over smooth surfaces the skin may become erythematous and stretched and tender The patients are often partially disabled by weakness and apprehension

The disease is not an established entity in regard to etiology or pathology It may last a long time and during this time there is a gradual increase in deposits of fat and in the accompanying discomforts

Fibromas A fibroma is a circumscribed purposeless arrangement of fibrous cells Fibromas are variable in size and consistency some are soft and loose in texture others are firm and hard to the palpating hand

Fibromas may occur anywhere in the body They arise in the skin fascia and intermuscular tissues about joints and in connection with nerves They may be situated in the ovaries breasts and uterus

Fibromas of the skin are often multiple hard and feel as though they were of subcutaneous origin These tumors may hang as if on a stalk from the labia or anal margins (see Recklinghausen s Disease in Chapter 6)

Mycosis Fungoides This is a rare condition seen in persons more than 40 years of age It is not a mycotic infection There is a scaly and extremely itchy eruption The disease after months or years may form tumors which generally ulcerate and form a fungating mass These scaly itchy lesions and finally ulcerating masses may affect the mucous membranes of the throat and intestines as well as all parts of the exterior surfaces The prognosis is grave

DISEASES AND INJURIES OF THE BREASTS

Development The earliest evidence of the development of the mammary gland is an epidermal ridge extending from the axillary region to the groin Certain areas in this line thicken and form the mammary glands This line is of interest chiefly because when accessory breasts form they are situated somewhere along its course

When accessory mammary glands persist most commonly it is the upper primary pair situated in the axillary region just over or beneath the edge of the lateral border of the pectoralis major muscle These may share in the functional activity of the normally situated mammary glands Accessory breasts may be observed first in the latter months of pregnancy when they prepare to lactate In rare instances the accessory glands may become painful just prior to menstruation when the normally situated glands are free from pain

The accessory mammary glands found in the axillary folds are of sufficiently frequent occurrence to make it necessary to remember them when considering the diseases of this region In instances in which there is a nipple associated with these accessory breasts any disturbance here will be self evident In those instances in which there is no nipple diagnostic difficulties may be encountered

Normally developed mammary glands which will give milk may develop outside of the mammary line Such glands may occur on the thigh buttocks or abdomen Accessory nipples (hyperthelia) without associated breast tissue may be found in the pectoral region and down the lateral aspects of the abdomen

Anomalies of Growth at Puberty Frequently when the breasts are stimulated to growth at puberty one responds at an earlier date than its mate and attains a considerable size while the other still retains the infantile state The importance of recognizing this condition lies wholly in the fact that the gland first to develop is normal and not malignant

Proof that the gland first developed is normal occurs when the previously undeveloped gland takes on growth and soon catches up with the other Why they may not simultaneously respond to the growth stimuli is not known

There is a sizable number of women who mature and bear children and who have well developed nipples under which there is scarcely any breast tissue These women have a tendency toward an excessive amount of coarse hair on the face around the nipples in the pubic region and on the thighs Their sexual desires are normal or excessive

Anomalies of Growth in the Mature Gland The mammary glands of different women vary greatly in size Why some mammary glands grow larger than others appears to be a matter of heredity Almost any size may be considered normal so long as the glands develop reasonably symmetrically Once their growth has ceased as after the development of puberty they may begin to grow again a condition termed hyperplasia or *hypertrophy* Whether the enlargement is of fat or gland tissue is frequently difficult to determine In many of these cases the disturbances are esthetic only and depend on the viewpoint of the patient The aging matron will tolerate with equanimity degrees of growth that would excite consternation in the maid In some cases the breasts become definite encumbrances Instances have been reported in which the breasts reached the thighs The development usually remains within the bounds of tolerance particularly if the patient has attained advanced years Less commonly one breast only takes on excessive development Such a breast is soft and compressible without any evidence of the presence of a tumor When the hyperplasia involves gland tissues the breast may be not only larger but notably firmer

Often the overgrowth is due to an excessive development of fat In these fat breasts the glandular elements are sparse and will not perform their natural function if the individual undergoes the puerperium In puerperium there is pain but no milk

Instead of producing disturbance at lactation periods some overgrown breasts do so during the general disturbances of the menopause Less commonly the fibrous tissue of an overgrown breast takes on elephantiasis like development In these cases the epithelial elements may also undergo hyperplasia and may give milk even in the virgin

THE ADULT MAMMARY GLAND

The breasts gradually develop until they attain the form which they are destined to assume in adult life varying much because of heredity When they first reach their adult form they are usually cone shaped carrying the nipples on their summits The breasts of some women maintain this form permanently but usually when complete maturity is reached they become more or less pendent varying much according to the nutritional state the individual familial and racial characteristics involved The actual and relative size is subject to wide variations of no clinical significance

When the adult breast is viewed in the sitting patient it is in most instances somewhat pendent At the apex the nipple protrudes and about it is the areola from which spreads in all directions the contour of the breast proper Thus the relations of the significant anatomic parts of the breast may be enumerated Each part now will be considered separately

The Nipple and Nipple Discharges The external variations of the nipple are considerable The nipple may protrude but little or may appear erectile even in repose In the average instance it is a truncated cone the surface of which is flat tened marked usually by pits or rugae or even a multitude of minute wartlike pro tuberances The nipple is capable of spontaneous elongation with puckering of the skin of the areola due to contraction of the circular nonstriated muscle fibers (erectile tissue) which surround the tubules The color of the nipple varies with the complexion of the individual In the blonde it is pale pink and in the brunette dark pigmented The color of the nipple and its environs usually become darker during the first pregnancy a shade which in a measure is permanently retained

The appearance of the nipple depends somewhat on the position of the patient during the examination A nipple that is normally protuberant in the recumbent posi tion may seem to be retracted particularly in pendent breasts when the patient is sitting *In some women nipples that appear retracted in the nonpregnant state* may become normal in pregnancy In others they may remain retracted

The *nipple discharges colostrum* a short time before parturition (colostrum gravidarum) Regularly after parturition colostrum (colostrum puerperarum) can be expressed from the nipple In some multiparous women colostrum can be ex pressed from the nipple between pregnancies In all other instances discharges from the nipples are significant

Benign breast disease which includes chronic cystic mastitis fibroadenoma and papilloma is the source of discharge from the breast in 3 of every 5 cases whereas malignant breast disease which includes Paget s disease is the source of discharge in the fourth case No sign of a palpable breast tumor can be found in the fifth of either benign or malignant cases

No one type of *nipple discharge* is specific for a particular type of breast lesion and conversely cancer as well as benign breast disease can give rise to diverse types of nipple discharge However in all who have papilloma and most of those who have blue dome cysts there is either a serous or a serosanguineous secretion from the nipple No diagnostic method is available which will localize these tumors if they are not palpable

Despite these facts no one can afford to take the responsibility for pursuing the policy of watchful waiting without surgical exploration when a bloody discharge from the nipple is present

Scattered over the extent of the *areola* in a circular fashion are prominent elevations due to the presence of the glandules resembling the racemose glands in structure In pregnancy they exude an oily liquid which should not be considered as a discharge from the nipple

The Gland The breasts present a variety of forms which vary with the position of the individual A cone shaped breast in the recumbent position is slightly pendent in the sitting posture This form being the average is regarded as the normal after young girlhood has been passed

The breast is composed of a series of from 16 to 20 primary ducts radiating from the nipple downward and outward After forming the ampullae the ducts then radiate toward the periphery of the breast The ampullae in turn end in lobulated cords varying much in size and complexity Surrounding the lobules are connective tissue septa which extend to the surface skin forming all together a support for the gland lobules These are the ligaments of Cooper They are important for their contraction causes the dimpling of the skin in malignant disease and they determine the size and shape of superficial abscesses or the pointing of deep ones

Most breasts are made up almost entirely of connective tissue with a sparse amount of fat The termination about the border may be gradual the fibrous tissue being interspersed with lobules of fat but the borders may be uniform and terminate abruptly Normal breasts may be so thick that when they are caught between the thumb and fingers they give the sensation of a heavy elastic pad Palpated with the flat hand this gives the impression of a flat soft tumorous mass suggesting a soft fibrosis On the contrary breasts may contain but little fibrous tissue it being limited to little more than septa Such breasts when palpated give the sensation of their main constituent fat The difference in palpatory findings in women of various ages is due to the changes in the character of the connective tissue rather than in its amount

The Lactating Gland The changes in the breasts incident to pregnancy result from the increase in size due to the enormous development of the acini the edema of the connective tissue and the increase in the circulation

In some cases the new acini may develop beyond the fibrous regions similarly to the hyperplasia seen in the lower animals This distribution of newly formed acini may be irregular abundant in some areas sparse in others In others even in the late stages of pregnancy the acinar hyperplasia may be confined to the fibrous regions In the first months of pregnancy the breast shows congestion and the superficial veins become dilated to such a degree that they form one of the early signs of pregnancy This is true at a time when there is little gross change in the gland itself

The Involutional Breast The mammary gland undergoes recessive changes after the menopause If these changes are orderly there is usually a general atrophy of the acinar and ductal epithelium and with this a lessening in amount and elasticity of the fibrous tissue In some breasts the atrophy of the acinar cells is preceded by a hyperplasia such as occurs in the mucosa of the uterus When this hyperplasia develops it should no longer be classed as a normal involutional change In most instances the hyperplasia subsides and the breasts then resume a normal recessive course In a certain number of cases in which the hyperplasia persists beyond the menopause the condition terminates in malignant disease

The topographic changes of the breast may be marked at or near the menopause because they respond to the physical change of the individual Many women gain much in weight at this time and the breasts too gain fat However as the emaciation of old age supervenes the breasts share in the decline until only a flabby bag of skin and fat remains

Gynecomastia The term gynecomastia refers to an enlargement of the mammary gland in the male When both breasts swell there may be an endo

crinologic cause such as neoplasm of the testicle or the administration of estrogenic hormone

Gynecomastia was present in 5 soldiers in one group of approximately 20 000 troops (Sullivan and Munslow) In none of the 5 men was there an endocrine disease present nor were there any associated endocrine disturbances except that the condition had developed in 4 during adolescence

More commonly the enlargement is due to inflammatory changes of a chronic type or to cystic disease somewhat similar microscopically to chronic cystic mastitis in the female If only one breast is affected a suspicion of neoplasm is aroused Symptoms may be absent or may consist of slight pain or discomfort The unilateral tumors should be excised even if small particularly because of the danger of cancer which may occur in the male breast

Mastodynia The term mastodynia connotes pain in the breast which may be severe enough to be completely disabling This symptom is rarely due to an associated chronic cystic mastitis Palpation of the breast reveals no abnormality This symptom has been designated as neuralgia of the breast and thought to be similar to the true neuralgias Treatment is unsatisfactory mammectomy may not relieve the pain in the region of the breast

INFECTIONS OF THE BREAST

Both acute and chronic inflammations of the breast occur

Mastitis is classified as interstitial and parenchymatous Both types are usually preceded by cracking or fissure of the nipple or areola which admits microorganisms to the subcutaneous lymphatics Often the fissuring and invasion are the end of the process and if so the mastitis is interstitial

Parenchymatous mastitis refers to those infections which are situated in the glandular and ductal portions of the breast The parenchymatous infections usually appear in the puerperium from the fifth or sixth day onward In marked contrast is the interstitial type which is common in the first days of the puerperium Either variety may end in suppuration with the formation of abscesses in various parts of the breast necessitating drainage

An occasional instance of mastitis is observed in the virginal or nonlactating breast during the course of some such disease as epidemic parotitis or after trauma

Acute Mastitis Formerly it was thought that *Staphylococcus albus* was the cause of acute mastitis More recently however it has been learned that *Staphylococcus aureus* is by far the commoner cause

Mastitis is always the result of infection Either pathogenic bacteria from outside gain access to the breast through fissured nipples by way of the lymphatics or some of the bacteria already present in the lactiferous ducts meet with conditions which enable them to invade the tissues

The symptoms of mastitis rarely appear before the end of the first week of the puerperium and as a rule not until considerably later Marked engorgement usually precedes the inflammation the first symptoms of which are chilly sensations or an actual rigor which is soon followed by a considerable rise in temperature and an increase in the rate of the pulse The breast becomes hard its surface is reddened and the patient complains of acute pain In many instances by the end of 24 hours the condition disappears spontaneously without treatment if the symptoms persist for longer than 48 hours suppuration is to be expected

Abscess formation often causes pronounced constitutional symptoms with or without severe local symptoms In still another group of cases the process pursues a subacute or almost chronic course the breast being somewhat harder than usual and more or less painful but constitutional symptoms are either lacking or very slight Under such circumstances the first indication of the true state of affairs is

often afforded by the detection of fluctuation on examination of a caked breast (stagnation mastitis)

On examination the breast is swollen, tense reddened and tender when moved The pain and local tenderness indicate the situation of the acute process When an abscess is present the process may be limited to a single quadrant If the abscess is not lanced the breast is likely to become undermined in all directions and as a result the destruction of tissue is extensive and numerous fistulous tracts form

The diagnosis of acute mastitis is obvious when there is a swollen hot reddened tender and painful breast

Chronic Mastitis Chronic mastitis may originate in the breast during the puerperium and persist for a long time The disease is often the sequela of abscess of the breast which has been poorly drained and in which fistulization is present

Soon after the onset of puberty a diffuse enlargement of the breast occurs If the breasts be traumatized by handling or squeezing and stimulated by sexual excitement a congestive condition may be initiated This congestion may pass on to a true inflammation and suppuration

On examination the breast is somewhat enlarged and contains one or more irregular areas which are firm and tender In the absence of a history of trauma in young women these areas should be biopsied for histologic study

Small localized alveolar abscesses occur in virgins For a time there may be localized redness of the skin milk pain and soreness These abscesses usually resolve spontaneously

Tuberculosis Tuberculosis of the breast may be the cause of persistent fistulas which have originated from what appeared to be an acute mastitis abscess formation and surgical drainage The first intimation that a tuberculous infection is present is often a failure to heal of the stab wound from the lancing of the abscess during lactation

A certain number of instances of chronic mastitis and particularly of chronic abscess formation are of tuberculous origin These instances in which the onset is a slow enlargement in a quadrant of the breast require separate consideration Tuberculosis of the breast occurs probably once for every hundred cases of carcinoma Pregnancy and lactation are thought to be predisposing factors It is usually unilateral and rarely exists in association with carcinoma

The disease begins as a painless irregular swelling in the breast The skin becomes reddened There are no systemic reactions from the breast Any systemic reactions are from a more profound tuberculous infection for instance in the lungs

The breast is swollen and slightly tender or there may be no tenderness and but little swelling A lump or lumps are present The nipple may be retracted If abscesses have formed the periphery of the tumor is hard its center soft The axillary lymph nodes may be enlarged on the side of the affected breast Often nodes in both axillae are palpable

The diagnosis is based on biopsy of the breast tissue Without fistulization this form of tuberculosis of the breast is differentiated from carcinoma only by histologic study Tuberculosis of the breast is usually primary develops by direct invasion and usually involves the regional lymph nodes The disease does not undergo regression has no complications and is treated successfully by radical mastectomy Drainage from a sinus tract should be submitted to microscopic and cultural studies for tubercle bacilli *Often there exists an underlying pulmonary tuberculosis*

Syphilis Gummas of the breast are rare They appear as isolated tumors but are more frequently multiple and bilateral Gummas may be subcutaneous but more commonly are intermammary They can be diagnosed only by means of biopsy studies

TRAUMATIC MASTITIS

(Fat Necrosis)

Injuries of the breast are commonly followed by necrosis of the fat in the injured region This may continue as a solid reactive tumor terminating in resolu tion or the center may break down producing an aseptic abscess

The trauma is usually an acute one for instance the bite of a dog a horse or a sweetheart Falls on the breast and injuries from being thrown against the steering wheel of an automobile are also common examples Following the trauma there are soreness and tenderness In other instances there is no history of trauma despite the finding of fat necrosis on histologic study

On section the center of the lesion is whitish a color which contrasts with the yellow fat about When organization of the affected area is well in progress the whitish hue may be obscured by the process of fibrosis The center may have undergone liquefaction producing an irregular cavity of small dimensions compared with the bulk of the palpable tumor

The tumorous mass is continuous with its environment It is hard The skin is not freely movable over it and may be reddened and edematous There is no dimpling of the skin and the nipple is not retracted unless it was so before the disease began nor is it fixed The tumor does not have the firmness of cancer

The diagnosis is usually obvious from the history and the examination but it is not certain until biopsy study is made

PAGET S DISEASE OF THE BREAST

Paget s disease is defined as a malignant papillary dermatitis affecting the areola and the nipple Arising first as a superficial dermatitis the chronicity the feeling of thickening and the unilateral character are the chief points worthy of note The disease appears commonly after the menopause and may simulate an infective dermatitis It is often exceedingly difficult to distinguish between the two conditions On the whole the infective dermatitis is moister and not so circumscribed but above all it yields readily to treatment In Paget s disease the lesion is more definitely thickened and circumscribed and may show some retraction of the nipple When allowed to continue the condition spreads into the breast and subsequently to the lymph nodes of the axilla

Biopsy is performed in all cases of suspected Paget s disease of the nipple which does not clear up in a week or two after a program of cleanliness and protection of the breast

Paget s disease of the nipple is treated the same as any other carcinoma of the breast by radical mastectomy Since many of these tumors are slow to meta stasize the prognosis is particularly good in patients without obvious metastasis

BOWEN S DISEASE OF THE BREAST

Bowen s disease is characterized by the formation of a thick pinkish tubercle or papule covered by a thickened horned layer of skin In contrast to Paget s disease Bowen s disease occurs on any part of the mammary skin and when malignancy supervenes it is of the squamous epithelial type

BENIGN CYSTIC TUMORS OF THE BREAST

(Endocrine Disturbances)

Chronic Cystic Mastitis This disease is called also Schimmelbusch s disease chronic interstitial mastitis chronic lobular mastitis cobblestone breast nodular breast mastoplasia mastopathia cyclomastopathy and adenosis

Chronic cystic mastitis in spite of its designation is probably not an inflam matory disease at all but is secondary to hyperplastic changes instigated by endocrine

activity The cause of these changes in the breast is now generally considered be an abnormality in the cyclic activity of the female sex hormones An essen feature of this disorder is the failure of normal involution so that by repea stimulation at each menstrual cycle during pregnancy and perhaps also by sex excitement the hyperplastic changes outstrip the hypoplastic or involutio process

The most important pathologic changes consist of a hyperplasia of the acinar the ductal epithelium particularly the latter and a hypertrophy of the firm transluc hyaline tissue surrounding the ducts There may be varying degrees of round cell infil tion which is the only histologic feature which points to inflammation The hyperpl in the ducts may produce obstruction or may be accompanied by excessive secreti thus producing cysts In some instances the hyperplastic ductal epithelium takes form of papillary ingrowths into the lumen of the duct intraductal papilloma or the lumen of a cyst intracystic papilloma

Very few women who have chronic cystic mastitis ever consult a physician they do they complain of a lump or pain or of discharge from the nipple

The pain is usually slight and brings the patient to the physician only becaus has aroused a fear that such a symptom may be a sign of cancer even though lump may have been felt Actually of course pain is not a manifestation of can except in the terminal stages when ulceration and infection have occurred Inde severe pain is generally indicative of the presence of other lesions such as ac mastitis or abscess The pain in chronic cystic mastitis is rarely constant be worse at some period during the menstrual cycle usually just before or during flow In a few patients however the pain may be severe enough to be disabl and demand relief under such conditions it is called mastodynia (see p 522)

Discharge from the nipple in chronic cystic mastitis is relatively uncomm being encountered in less than 10 per cent of cases In a study made by Hicl the discharge was bloody in about half the cases and serous or greenish bro in the remainder In ductal papillomas the incidence of discharge is much hig and the excretion is usually bloody for example Geschickter noted a blo discharge in 47 per cent of 204 cases studied When ductal papillomas are in just beneath the nipple they may be palpable but when located in the depth the breast they are rarely palpable because of their small size A significantly la proportion of ductal papillomas undergo malignant change

In a breast which is the site of chronic cystic mastitis a diffuse nodular or sh sensation will be encountered on palpation of the parenchyma This will be p ticularly apparent in thin pendulous breasts in which palpation is not impeded a thick layer of subcutaneous fat Often the lateral margin of the breast parenchy will be felt as a sharp firm edge Both breasts are affected and often equally When a single lump is felt it is firmly attached to the rest of the breast parenchy and cannot be outlined as a separate tumor In many patients however the dif entiation between an actual tumor and a localized region of cystic mastitis may made only after excision On repeated examinations it will often be noted t the nodules vary in size and in the degree of tenderness at various periods du the menstrual cycle In general the signs are most prominent before and during flow though they are not infrequent in the midperiod between menstruations Th breasts are always somewhat tender on palpation The tenderness extends to pectoral muscle

The lump may be single but if moderate in size it is usually due to a la cyst more often the lump is rather small and in reality represents a portion o diffusely nodular breast which for some reason becomes locally more promin or nodular than the rest of the parenchyma In many cases however the pati believes she feels a lump but on examination only a diffuse nodular parenchy is made out

A definite diagnosis is made by biopsy and histologic study of one of the tumors

Single Cyst of the Breast Occasionally larger single cysts are encountered which are manifest clinically as lumps There usually are no other symptoms However in some instances serous or serosanguineous nipple discharges may be present On examination although the contents of the cyst are serous they are under so much pressure that they are as firm as a solid tumor, often imparting a hardness that arouses suspicion of carcinoma The walls of large cysts of this type present a bluish translucent appearance when exposed at operation the term blue dome cyst has been given to such a lesion by Bloodgood The diagnosis is made only by thoroughly examining numerous sections of the excised tissue

A *galactocele* is a true retention cyst, due to obstruction of a duct and contains retained milk It is relatively rare and has little clinical significance because it usually empties spontaneously If it does not empty the contents will become inspissated removal may then be indicated

BENIGN SOLID TUMORS OF THE BREAST

The breast may be the site of any of the tumors such as adenoma lipoma fibroma and angioma which are found in skin and subcutaneous tissue

Adenoma (Adenofibroma) Adenofibroma is a tumor of early adult life often appearing in girls just after puberty and in young women If the patient has known of the existence of the lump for a number of months or years she may notice a definite though slow increase in size Only rarely are the tumors multiple Rapid growth may occur during lactation owing to the same stimulus which affects the rest of the breast many women indeed first notice the lump during such a period

It is thought that this tumor may have a similar origin to that of the adenomas of the thyroid If this should be true adenomas of the breast may be classed not as neoplasms but as a result of endocrine stimulation

Adenomas contain more fibrous than epithelial elements The adenomatous tissue may be so slight as to form single layers of epithelial cells surrounded by great masses of connective tissue thus giving rise to the term intracanalicular fibroadenoma Mucoid degeneration may occur and then the tumor is designated adenomyxoma The micro scopic structure sometimes bears a resemblance to that of chronic cystic mastitis

Until a lump is discovered usually accidentally symptoms are absent Pain and nipple discharge are occasionally present In most instances the patient feels the mass while she is bathing or notices it on examining her breast after some slight trauma This latter fact accounts for the frequent belief on the part of patients that the tumor followed an injury to the breast Many women however now palpate their breasts regularly because of the lay propaganda conducted by the American Society for the Control of Cancer

On examination a firm tumor which is movable and distinct from the breast parenchyma is palpated The mass is usually larger and more definite than the lumps characteristic of chronic cystic mastitis

A definite diagnosis cannot be made with certainty on palpation alone A decision therefore must be made in each case as to the indications for surgical intervention When a breast is removed from a young woman she feels that a barrier has been placed between herself and a fundamental object in life Realization and adjustment for her always involve the deepest forces of her personality This is true not only of the young woman but even of those of advanced age years after the breast has performed whatever function it may or may not have been intended to perform

Operation is not indicated for a benign solid tumor of the breast if the tumor is not painful and is present in a young girl In general surgical excision is indicated in most women more than 25 years of age and in every woman more than 35 years

of age who has such a tumor Excision and microscopic examination permit of a correct diagnosis

Brodie s Disease of the Breast The term Brodie s tumor should be reserved for large excessively cellular and long neglected fibroadenomas The history of a pre existing breast tumor of many years duration which at first grows slowly or not at all and finally in recent months increases rapidly in size is the outstanding feature of Brodie s tumor The skin overlying the tumor is mobile and the lymph nodes are not usually involved Enlargement may be so great that ulceration of the skin occurs from pressure necrosis and the foul fungating mass suggests a carcinoma Sometimes the correct diagnosis of an innocent large tumor is impossible until the breast is removed and histologic studies are completed

Papillomas of the Duct A serosanguineous discharge from the nipple often is characteristic of a papilloma in the lacteal ducts The bleeding from the nipple may be unaccompanied by any other symptoms for a period of several years Bleeding may start from a slight trauma or spontaneously and it may continue daily with the passage of a few drops or intermittently every 1 or 2 months In some this symptom may become more persistent at the time of the menopause Papillomas of the duct may occur in aging men

On examination not more than one half of these patients will have a palpable tumor On histologic study of the cyst it may be found to be either benign or malignant

Lipoma Lipomas of the breast are completely benign tumors They cause trouble mainly from worry and anxiety of the patient because of their presence However no one can be sure that the tumor is a lipoma until biopsy studies are made

Lipomas occur in the same age groups as carcinomas Gynecomastia excluded lipomas are the commonest tumor of the male breast Lipomas do not originate in breast tissue They begin in the subcutaneous tissue above the breast or in the submammary tissue along the lower border of the pectoral muscle and displace the breast forward

Lipomas may be soft superficial and apparently situated on top of the mammary tissue They may be lobulated on palpation and may cause dimpling of the skin When situated along the lower border of the pectoral muscle their identity is less easily recognized

MALIGNANT TUMORS OF THE BREAST

Carcinoma of the breast is predominantly a disease of women (100 1) It comprises more than 90 per cent of all malignant tumors of this gland sarcoma comprises but 3 to 5 per cent

More than half the women who have carcinoma of the breast are more than 50 years of age Other than aging there is nothing known of the etiology Thus the relation of chronic cystic mastitis to cancer is not known However cancer develops in less than 1 per cent of the cases Experimental cancer has been produced by repeated injections of estrin the significance of this fact is not known The relation between skin irritation in Paget s and Bowen s diseases and cancer is more definitely significant Trauma and previous lactation have no proved etiologic connection with the disease

PATHOLOGY Cancer of the breast arises from the ductal epithelium or from acinous tissue The various types of carcinoma of the breast are classified according to the relative proportion of fibrous and epithelial elements the rapidity of growth and particularly the microscopic arrangement of the cancer cells for instance their tendency to form definite glandlike structures Thus three main types are described medullary carcinoma scirrhous carcinoma and adenocarcinoma

Medullary carcinoma is a cellular and in general a rapidly growing tumor It is soft and it is more frequently found in young women than in those of more advanced age

Scirrhous carcinoma is composed largely of fibrous tissue The large amount of fibrous tissue gives the tumor its characteristic stony hard sensation on palpation It about equals medullary cancer in frequency of occurrence but its victims are women of more advanced age It grows more slowly and tends to metastasize late

Adenocarcinoma derives its name from an arrangement of cells in the form of alveoli or acini These tumors vary in their rate of growth Some grow rapidly and metastasize widely Others are present for years before metastasis occurs The tumor may invade the skin early and produce extensive ulceration Colloid carcinoma is a relatively rare form of adenocarcinoma *in which much colloid material is present*

The extension of cancer of the breast is by direct permeation into small lymphatic channels of the mammary lobules and into the subcutaneous tissue fascia muscle and skin Once the tumor cells are in the lymphatic structures of the skin the *permeation is made evident by small nodules just beneath its surface This condition* is commonly a manifestation of local recurrence of a tumor after surgical removal has been attempted Tumors situated in the medial half of the breast involve the parasternal lymph nodes Extension along fascial lymphatic channels which penetrate the pectoral and the upper end of the rectus muscles and the thoracic wall is common

Metastasis through the larger lymph channels occurs to the regional axillary lymph nodes and occasionally to the supraclavicular nodes In many metastasis occurs by way of the blood stream to the liver and to the spinal column and long bones Metastasis to the liver may spread quickly and produce an abdominal carcinomatosis with all of its dire consequences In some instances of cancer of the breast metastasis will appear in the ovaries (Kruckenberg tumor)

Classification of Cancer of the Breast Surgeons have employed various criteria based on clinical or pathologic manifestations singly or combined in an effort to predict curability or incurability by surgical treatment The classification arranged by Portman is adapted for use here The indications for different thera peutic procedures for cancer of the breast and the general average of results as reported in the literature may be discussed on the basis of the classification and criteria of incurability presented

In Portman s classification four stages of the disease are recognized and set forth as follows

First Stage The earliest clinical sign of a cancer of the breast is a small freely movable tumor without skin involvement palpably enlarged lymph nodes or evidence of remote metastatic lesions There are other tumors of the breast presenting the same clinical signs which are not cancers therefore it often is impossible to make a *differen* tial diagnosis Microscopic examination is necessary

About 15 per cent of patients whose lesions have been considered to be of stage 1 after operation and microscopic examination die within 5 years The reasons probably are (1) *Neoplastic cells may not have been arrested in axillary nodes* but may have passed through them and metastasized elsewhere presenting no clinical or pathologic evidence prior to operation (2) The cancer may have metastasized not to the axilla but to the internal mammary or mediastinal groups of nodes as happens especially in the case of inner quadrant tumors (3) Metastatic lesions may not have been detected by microscopic examination

Second Stage The second stage in the progress of cancers of the breast usually embodies a movable tumor without skin involvement but with metastatic extension in a few axillary nodes none being demonstrable elsewhere The prognosis after operation is almost in direct ratio to *the relative number of nodes involved About* 25 per cent of patients who undergo operation will have only a few nodes involved (stage 2) and the general average of five year survivals has not been more than 50 per cent by operation alone

Since 50 *per cent* of stage 2 patients who [illegible] disease

within 5 years it must be because extension had taken place beyond the anatomic limits of surgical removal Although recurrences seldom take place locally in the chest wall or in the axilla of those who have undergone radical mastectomy the disease probably has extended to the infraclavicular or supraclavicular and parasternal nodes and eventually will spread to vital organs It would seem logical to give postoperative roentgen therapy in such cases in order to delay the progress of the residual cancer This will be accomplished in about 15 per cent of cases

Third Stage The third stage of cancer of the breast includes cases with metastatic lesions in many axillary nodes skin involvement (dimpling) and with or without other criteria of incurability In some cases in the latter classification the presence of metastatic *involvement in many axillary nodes* can be determined only after operation The prognosis is little if any better in such cases than in those with criteria of incurability However most cases in stage 3 will be classified as having these criteria

The general average of five year survivals by operation alone for stage 3 cases has been about 5 per cent It has been found that the average natural life expectancy of women with cancer of the breast who have received no treatment is almost 3 years and 5 per cent live 5 years Therefore it may be concluded that radical operations for stage 3 cases do not increase survival rates

Fourth Stage A fourth class of cases of cancer of the breast must be made to *include those with remote metastatic lesions on initial examination regardless of the* presence or absence of criteria of incurability A complete physical examination and a roentgenographic survey of the lungs skull vertebrae and pelvis should be made of all patients with tumors of the breast to exclude the possibility of metastatic lesions before therapeutic procedures are instituted especially when there is a suspicion of cancer Metastasis to these regions or other viscera may develop from cancers which appear to be clinically in early stages Patients with remote metastatic lesions are not wittingly submitted to operation and the majority are treated for palliation by irradiation or other measures such as endocrine therapy or by both In one series 20 per cent of patients *were found on initial examination to have remote metastatic lesions*

It should be noted that about 45 per cent of all patients observed by Portman were classed in stages 3 and 4 combined when first examined Such cases are incurable and the average of five year survivals after operations alone is no greater than if no operation were performed—perhaps less

EXAMINATION A carcinoma of the breast is a firm often hard tumor that cannot be separated from the surrounding breast tissue or moved within this tissue The contour the size the shape and the nipples of the breasts are compared An indentation or a bulge in the contour may reveal the site of a lesion The contour of a breast may be altered by the height of the two breasts on the chest wall and their relative size

Very commonly one breast is the larger of the two If the general contours of the two are the same the difference in size usually is normal being a developmental difference

The shape of the nipples and the axes in which they point are compared The surface of the nipples is inspected for evidence of crusting or erosion

The enlarging carcinoma fixes the fascial septa of the breast exerts an abnormal traction on them sometimes pulls the skin inward to produce dimpling and deviates the axis in which the nipple points or flattens and retracts the nipple If the carcinoma has extended sufficiently to be attached to the pectoral fascia the pectoral muscle may be contracted the sector of the breast in which the carcinoma lies is pulled upward abnormally and its contour is distorted The horizontal levels of the areola and nipple are often elevated by a carcinoma in the upper half of the breast

Deviation of the axis in which the nipple points is a subtle sign of retraction The fibrosis in and about the *carcinoma pulls* on the ductal system and tilts the nipple so that it points toward the carcinoma

When the carcinoma involves the area beneath the nipple the fibrosis shortens

the whole ductal system and pulls the nipple inward This process may reveal itself as flattening and broadening of the nipple on the side of the carcinoma As the fibrosis progresses the nipple becomes flatter and broader until in some instances it is finally retracted beneath the surface of the surrounding areola

Dimpling of the skin over a carcinoma of the breast may be demonstrated in some cases by the patient s raising her arms high above her head However, dimpling is most easily and uniformly demonstrated by gentle compression of the breast between the hands

The relative degree of fixation of the carcinomatous region in the breast to the pectoral fascia is best demonstrated by determining the amount of lateral excursion of the tumor while the patient lies supine by gently moving the tumorous portion transversely across the chest wall between the fingers of the examiner s two hands first with the patient s arm relaxed and then with her hand pressed against the hip to contract her pectoral muscles

Supraclavicular and Axillary Regions During the examination of the supra clavicular regions the patient sits facing the examiner The supraclavicular space is palpated with the ends of the fingers Nodes in this region are often softer than those in the axilla

In the examination of the axilla ask the patient to place her hands on top of her head while the lower limits of the axilla are examined Then have her lower her arms to her sides while the upper limits of the axilla are examined with the tips of the fingers The right axilla of the patient is examined with the left hand The left axilla is examined with the right hand Lymph nodes high up in the axilla are difficult or impossible to feel Nodes lying close behind the thick body of the pectoralis major muscle as it forms the ventral wall of the axillary space are also difficult to palpate In obese patients palpation of the axilla is particularly unreliable

The number consistency and movability of the axillary lymph nodes are observed The nodes may be fixed to the underlying axillary structures or to the overlying skin The transverse diameter of the largest axillary node is estimated in centimeters

Palpation is inaccurate in determining whether or not axillary lymph nodes contain metastatic lesions These nodes may enlarge from causes other than malignant disease

Diagnosis The diagnosis of cancer of the breast is often obvious from the presence of one or more of the foregoing findings If these findings are definite the carcinoma has reached an advanced stage It is desirable to find a tumor in the breast so small that biopsy is indicated for making a positive diagnosis Histologic study is the certain way of making a correct diagnosis

Early in the course of cancer of the breast metastasis tends to take place to the cervical and axillary lymph nodes ribs and spinal column liver pelvis and abdominal viscera and the skin

Bilateral Cancer of the Breasts Bilateral cancer of the breasts occurs frequently enough so that the presence of a known cancer of one breast requires study of the other breast particularly if there is a lump in the second breast Bilateral carcinoma occurring simultaneously is rarely found but it does happen The carcinoma of the remaining or second breast may occur years after the first one has been removed Often it is difficult for the pathologist to be positive whether the malignant lesion in the second breast is primary there or secondary or metastatic from the first breast involved

Inflammatory Carcinoma of the Breast Inflammatory carcinoma of the breast implies the coexistence of carcinoma and inflammation The fact is, however that these breasts usually do not contain evidence of inflammation on histologic study The condition often occurs in lactating breasts but this incidence is by no means constant

Pain is a commoner first symptom in the so-called inflammatory carcinoma of the breast than in the usual instance of carcinoma

Physical examination reveals diffuse induration extending through much or all of the breast So dense is the inflammation that often a definite tumor cannot be palpated

Inflammatory carcinoma frequently enlarges the breast Enlargement of one breast may be the first intimation of trouble The skin of the breasts is red and often edematous Axillary lymph nodes are always palpable Despite extensive involvement of the breasts ulceration of the skin is rare When present it occurs very late in the course of the disease All studies on inflammatory carcinoma of the breast indicate a poor prognosis

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Part Two

The Systemic Diseases

In medicine the term system has one of three meanings (1) a set or series of parts or organs which unite in a common function (2) a whole bodily organism which unites in a common function and (3) a school or method of practice

In this part of this book the first meaning is applied Hence the term systemic disease refers to disease affecting one or all parts which are related to perform a certain function for instance disease of the lungs trachea bronchi and pleura one or all of them is disease of the respiratory system

9

DISEASES OF THE THORACIC WALL, PLEURAE, MEDIASTINUM, BRONCHI AND LUNGS

THE THORACIC WALL

The lungs and heart may be hampered in the performance of their functions by diseases of their own as well as by being influenced by diseases of the body as a whole Hence the condition of these organs often serves as a guide to the general bodily functioning and therefore the state of the respiration and circulation is continually being examined for purposes of diagnosis prognosis and treatment It is for these reasons that the physical examination of the thoracic organs occupies a prominent place in diagnosis by physical means Once a physician has gained even moderate skill in examination of the thorax he has gone a long way on the road of rendering good service to patients These statements do not imply that an examination by physical means is all that is necessary in diagnosing diseases of the thorax They simply mean that examination of the thorax by physical means is always necessary At times the circumstances may be such that examinations by any other means are not feasible and then examinations by physical means may have to suffice for the basis of treatment

Shape of the Thorax The bony chest is subject to defects in development as well as to disease and injury and to deformities due to these causes

The thorax is conical in shape being small above and large below In infancy the anteroposterior and the transverse diameters are nearly equal At the age of 2 years the chest has become oval and in adults the transverse diameter is one fourth greater than the anteroposterior

Variations in the shape of the thorax are mainly the result of disease In childhood rachitic disease (rickets) produces a lateral flattening and a projection of the sternum If the sternum projects markedly it constitutes what is known as pigeon breast the thorax in such a condition being longer from front to back than from side to side In this disease also there may be a depression on each side of the sternum the back is rounded owing to the bending of the vertebral column and the points of junction of the ribs and cartilages are enlarged these enlarged points constituting what is known as beading of the ribs This line of beads is called the *rachitic rosary* From the level of the ensiform cartilage a groove passes out toward the sides this is known as *Harrison s groove* Some times the lower end of the sternum is pressed inward forming a deep funnel shaped depression constituting the deformity known as *funnel chest* This shape of the chest may be produced in children by obstruction to the breathing from enlargement of the tonsils from the presence of adenoid growth in the pharynx and from hypertrophy of the

turbinate bones all of which obstructions interfere particularly with nasal respiration

Diseases of the lungs and pleurae alter the shape of the thorax These common alterations are considered in the section on examination of the lungs and pleurae

Topographic Examination of the Thorax On inspection of the chest it may appear normal rounded or flat One side may be larger than the other and the intercostal spaces may be obliterated or retracted The gross abnormalities are observed and recorded

The clavicles and the sternum are subcutaneous and can readily be felt beneath the skin The point of junction of the first and second pieces of the sternum is opposite the second costal cartilage It forms a distinct prominence the angulus sterni or angle of Louis

The tip of the xiphoid or ensiform cartilage can be felt even in the obese patient The top of the sternum is opposite the lower edge of the second thoracic vertebra The angulus sterni is opposite the fifth thoracic vertebra The tip of the ensiform cartilage is opposite the eleventh thoracic vertebra Above the upper end of the sternum is the *suprasternal notch* or depression, below its lower end is the infrasternal depression or epigastric fossa

The *sternoclavicular joint* possesses an *interarticular cartilage* between the clavicle and the sternum which separates them sufficiently to allow the formation of a distinct depression which can be felt Below the inner end of the clavicle the first rib can often be seen and felt

The cartilage of the seventh rib is the last to articulate with the sternum The tenth rib is the lowest which is attached anteriorly the eleventh and twelfth being shorter and floating ribs The intercostal spaces are wider anteriorly than posteriorly and the third is the widest

In men the nipple is usually in the fourth interspace or on the lower border of the fourth rib and on a line a little to the outer side of the middle of the clavicle In women the mammary gland reaches from the third to the seventh rib

The operation of paracentesis or tapping for pleural effusion is often done in the sixth interspace in the midaxillary line or posteriorly (see Back in Locomotion, Chapter 6)

Directional Terms and Topography of the Thorax and the Respiratory Organs For the sake of convenience in description and record the thorax arbitrarily is divided into various regions and marked by longitudinal lines

The Longitudinal Lines Seven longitudinal lines are used They run parallel with the long axis of the body Three are situated anteriorly (Fig 9 1) and two posteriorly

1 The *median line* is the midline of the body This runs down the middle of the sternum anteriorly and the middle of the back posteriorly

2 The *parasternal line* runs parallel to the edge of the sternum and midway between it and the midclavicular line

3 The *midclavicular line* also called the *mammary line* is a longitudinal line passing through the middle of the clavicle This usually passes $\frac{2}{5}$ to $\frac{4}{5}$ inch (about 1 to 2 cm) medial to the nipple

4 The *anterior axillary line* passes through the anterior fold of the axilla

5 The *midaxillary line* passes through the middle of the axilla

6 The *posterior axillary line* passes through the posterior fold of the axilla

7 The *scapular line* passes longitudinally through the lower angle of the scapula

The Regions of the Thorax In the middle of the surface of the thorax anteriorly there are three regions

1 The *suprasternal region* is the part above the sternum between the sternocleidomastoid muscles It is the suprasternal notch

2 The *upper sternal region* extends from the suprasternal notch to a line drawn opposite the third costal cartilages

3 The *lower sternal region* is behind the second piece of the sternum from the third costal cartilages down

Anteriorly but *laterally* to the middle regions of the thorax there are four regions

1 The *supraclavicular region* above the clavicle This includes the supraclavicular fossa

2 The *infraclavicular region* below the clavicle down to the upper edge of the third rib

3 The *mammary region* from the upper edge of the third to the upper margin of the sixth rib This extends laterally from the edge of the sternum to the anterior axillary fold and has the nipple nearly in its center

4 The *inframammary region* extends from the upper margin of the sixth rib to the lower margin of the thorax

Laterally on the thorax between the folds of the axilla there are two regions

1 The *upper axillary region* extends down to the upper border of the sixth rib

2 The *lower axillary region* extends from the upper border of the sixth rib to the lower edge of the thorax

Posteriorly there are four scapular regions

1 The *suprascapular region* is above the spine of the scapula

2 The *scapular region* is the part covered by the body of the scapula below its spine

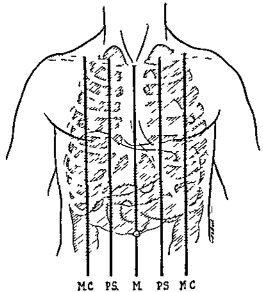


Fig 9 1 Topographic areas of the anterior thorax M C midclavicular P S parasternal M median or midsternal When used designate right or left side of patient

3 The *infrascapular region* is the part of the thorax below the scapula between its angle and the lower edge of the chest

4 The *interscapular region* is the part between the posterior edge of the scapula and the median line

The Pleurae These form closed spaces which line the thorax (parietal pleura) and cover the surface of the lungs (visceral pleura) The pleurae are completely in contact with the lungs only when the lungs are fully expanded In ordinary breathing the lungs are not completely expanded hence the edges of the pleurae fall together and so prevent the formation of a cavity This collapsing of the pleurae takes place mainly along the anterior and lower borders of the lungs

The apex of each pleura is attached to the tip of the cervical fascia There is an attachment to the first rib and to the scalenus anticus muscle which prevents collapse (Fig 9 2)

The upward extension of the pleura is about on a plane with the superior surface of the first rib The posterior portion of the pleura is higher than its anterior portion at the anterior end of the first rib The pleura extends 1 inch (about 2 5 cm) above the level of the superior surface of the clavicle

The top surface of the pleura is stretched in the form of a plane which is almost on a level with the superior surface of the first rib From the top the pleura slopes forward behind the sternoclavicular joint to meet the pleura of the opposite side at the level of the second costal cartilage The pleura covering each lung then descends to opposite or a little below the fourth costal cartilage where it diverges toward the side thence to the upper border of the seventh costal cartilage near its sternal junction thence downward and outward to the lower border of the seventh rib in the mammary line the ninth rib in the axillary line and the twelfth rib posteriorly The scapular line intersects the lower edge of the pleura at the eleventh rib

The Lungs When the lungs distend they expand mostly downward but also to a lesser extent laterally and anteroposteriorly owing to the elevation of the ribs which is due to the traction of the muscles upon them Breathing is performed mainly by the inter costal muscles and diaphragm As the diaphragm descends the air is drawn into the

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are not due so much to a bulging or to a retraction of the lungs as to the atrophy of the fatty and muscular tissue in tuberculosis and to the muscular tension in emphysema

In coughing the apex of the lung does not bulge into the neck above the clavicle as it appears to do but remains below the plane of the top of the first rib The appearance of bulging is caused by the movements of the trachea in the median line and of the muscles laterally This is noticeable particularly in the platysma and omohyoid muscles In quiet breathing the posterior belly of the omohyoid lies about level with the clavicle *but in coughing it rises 1 or 2 cm above it The intercostal membranes and* muscles are kept tense by the constant elevation of the ribs due to the muscular tension

OUTLINE OF THE LUNGS *Apex* The apex of each lung has its highest point opposite the posterior extremity of the first rib (Fig 9 2) It then follows the plane of the top of the first rib down to the sternoclavicular joint immediately above the junction of the cartilage of the first rib with the sternum The anterior end of the first rib is about 2 inches (5 cm) lower than the posterior The upper edge of the clavicle is 1 inch (about 2 5 cm) above the anterior end of the first rib and 2 5 cm below the head of the first rib hence the apex of the lung rises 1 inch (about 2 5 cm) above the clavicle and it lies behind its inner fourth This distance will vary in different individuals with the obliquity of the ribs The more oblique the ribs the greater will be the distance between the level of the top of the clavicle and that of the neck of the first rib The top of the lung however never is above the level of the neck of the first rib There is no appreciable difference in the height of the right lung and the left lung (Fig 9 3)

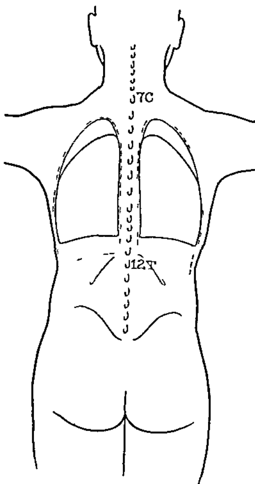


Fig 9 3 Outlines of the lungs—posterior view

Anterior Border From the sternoclavicular joint the borders of the lungs pass downward and inward until they touch or almost touch in the median line at the angle of Ludwig opposite the second costal cartilage Below this the right lung extends a little across the median line and the left recedes slightly away from it The right lung leaves the sternum opposite the sixth costal cartilage to which it has gradually descended

The left lung on reaching the level of the fourth costal cartilage curves outward and downward across the fourth interspace to a point about 1 inch (2 5 cm) to the inner side of the nipple in the fourth interspace From this point it goes downward and inward across the fifth rib and interspace to the top of the sixth rib about 1⅕ inches (3 cm) to the inner side of the nipple line This isolated tip of lung just above the sixth rib over the apex beat of the heart is called the lingula

Lower Border The lower edge of the lung varies in different individuals and in the same individual according to the amount of inflation In quiet respiration it

trachea and lungs and the pressure within the thorax is lowered to zero or to a minus quantity so that if it were not for its bony framework the chest would collapse The framework is sufficiently strong to retain its shape despite this pressure if the breathing is normal and the thoracic walls are healthy When however obstruction of the air passages is present in young patients the deformities funnel breast pigeon breast or variations of

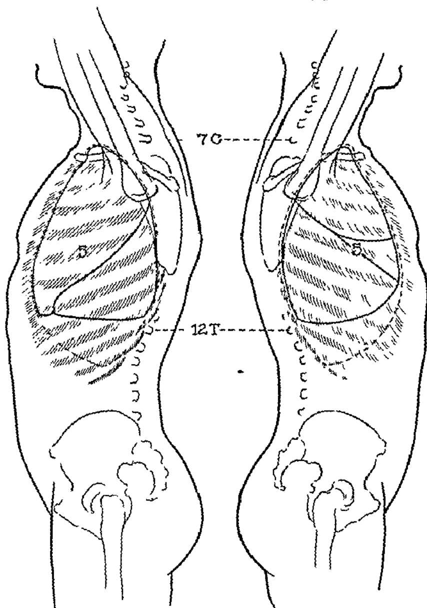


Fig 9 2 Lateral views of the extent of the pleura and outline of the lobes of the lungs

these are produced These deformities are also produced by weaknesses in the bony thorax such as occur in rickets

Enlargement of the chest posteriorly is impossible because of the support of the ribs vertebrae and strong back muscles Enlargement downward is allowed by a descent of the diaphragm hence the fullness of the abdomen in those affected with emphysema and conversely the flatness of the abdomen in those whose lungs are contracted In the region of the apices the thorax is closed by the deep fascia which spreads from the trachea esophagus muscles and great vessels and blends with the pleura to be attached to the first rib In disease this level with the plane of the first rib is not materially altered The apparent fullness of the supraclavicular fossae and intercostal spaces in emphysema and the increased depth of these hollows in pulmonary tube ulosis

tumor a single myeloma and giant cell tumor must be considered and when multiple regions are seen metastatic malignant disease or multiple myelomas are possible

Malignant tumors of the sternum may arise in the sternum and its neighboring cartilage They grow posteriorly encroaching on the anterior mediastinum Likewise tumors may arise from the spinal column and its surrounding structures and extend anteriorly and encroach on the posterior mediastinum They may extend laterally displacing the lung and pleura and occupying the apical or other thoracic regions At first from either point of origin these tumors remain circumscribed Later they extend through their own capsule and invade or surround the mediastinal structures These tumors may extend to the pleura and lungs and may metastasize to the lungs liver spleen kidneys and other organs

The symptoms of malignant tumors of the sternum are pain cough dyspnea tumefaction and mediastinal pressure These symptoms develop and progress more slowly than when lymphosarcomas are present in the mediastinum and producing similar symptoms

Physical examination may reveal a mass near the site of origin especially if the tumor arises from the sternum or its cartilaginous attachments Any or all of the physical signs of mediastinal tumors with compression phenomena may be elicited

Both anteroposterior and lateral roentgenograms are useful in demonstrating the size and situation of the tumor At times the roentgenographic study may be helpful in establishing the nature and origin of the growth The presence of hemorrhagic pleural fluid may indicate the presence of malignancy in the thoracic cavity

Surgical removal in the early stages is the preferred treatment

SYMPTOMS OF RESPIRATORY DISEASE

The symptoms often significant of pulmonary disease are pain in the chest cough expectoration hemoptysis cyanosis dyspnea stridor and hoarseness

PAIN IN THE CHEST Pulmonary tissue and the visceral pleura are devoid of the sense of pain The parietal pleura is richly supplied with sensory nerves from the intercostals Stimulation or irritation of the parietal pleura produces a sharp pain which can be located with fair accuracy over or near the point of stimulation The diaphragmatic pleura is also sensitive but stimulation of it never produces local pain the pain is always referred The central part of the diaphragm is innervated by the phrenic nerve and the afferents from here enter mainly the third and fourth cervical posterior nerve roots the peripheral part of the diaphragm is innervated by the lower intercostals and the afferents enter the seventh to twelfth posterior nerve roots Pain therefore from the central portion of the diaphragm is referred to the neck and the upper part of the shoulder from the outer parts of the diaphragm it is referred to the lower part of the thorax lumbar region and abdomen The stronger the pleural stimulation the wider is the area of the referred pain and it may even extend to the other side of the body

There are many types of pain which may be felt in or about the thorax all of which are commoner than those arising in the pleurae and lungs For instance there are muscular pains due to strain or even tearing of muscle fibers the boring pain of an eroding aneurysm root pains attending tabes dorsalis syphilitic meningitis tumors of the cord herpes zoster spondylitis and other lesions which involve the posterior nerve roots the substernal pain on swallowing which may be associated with esophageal obstruction There are the pains of angina pectoris and aortitis Pains in the breast may be considered to be pains in the chest Referred pains from perforating peptic ulcers are by no means uncommon The pains of true intercostal neuritis are common but difficult to diagnose

is about opposite the sixth cartilage and rib from the sternum to the mammary line opposite the eighth in the midaxillary line the tenth in the scapular line and the eleventh near the vertebrae The lowest part of the lung is on the side at the midaxillary line or a little posterior to this but the line from here posteriorly although rising slightly is nearly horizontal

THE FISSURES AND LOBES OF THE LUNGS The left lung has one fissure and two lobes an upper and a lower The right lung has two fissures and three lobes an upper a middle and a lower (Fig 9 2)

The relations of the fissures to the surface are variable They generally appear posteriorly at different levels the right main fissure being lower than the left The fissure of the left lung appears at the side of the vertebral column at a variable point extending from the third rib to the upper border of the fifth rib In the axillary line it is at the fifth rib although it may be slightly higher or lower and it ends beneath the sixth rib or the interspace above or below anywhere from 2⅔ to 4⅓ inches (about 6 to 11 cm) from the midline

The main fissure of the right lung leaves the vertebral column opposite the fifth rib or at the root of the spine of the scapula The fissure tends to follow the fifth rib being beneath the rib or beneath its interspace in the midaxillary line and ending at the sixth rib in the mammary line

The subsidiary fissure of the right lung leaves the main fissure in the posterior axillary line opposite the fifth rib and running horizontally forward ends near the sternum at any point between the third and fourth interspaces

Diseases of the Thoracic Wall The diseases of the thoracic wall are usually secondary to those primarily affecting the skin and subcutaneous tissues muscles bones and nerves or to diseases originating from the structures within the thoracic cage

Tumors of the Thoracic Wall Both benign and malignant tumors occur in the thoracic wall Lipomas are of common occurrence over the shoulders and back and in the axilla Likewise fibromas occur in the same situations Cysts may arise from the cutaneous glands over the back Pigmented nevi in aging persons are of common occurrence over the back

The primary malignant tumors of the thoracic wall arise in the skin subcutaneous tissues the breast and the bony framework The malignant tumors of the *skin* are those common to glabrous skin They are carcinomas and melanoepitheliomas Carcinomas when present are often situated over the shoulders and scapular regions originating in sebaceous glands which abound here The tumors of the *soft tissues* which undergo malignant change are fibromas and hemangiomas These tumors are described in the section on diseases of the skin A common malignant tumor of the thoracic wall is carcinoma of the breast The tumors which originate in the *bony framework* are multiple myelomas Ewing s tumors of the ribs osteogenic growths and chondrosarcomas all of which are rare and are considered in this text with tumors of the bones

Metastatic tumors of the ribs are frequent They may be clinically silent causing no symptoms or at times they may be the first manifestation of malignant disease when a pathologic fracture occurs Malignant tumors of the breast prostate gland and thyroid gland and hypernephromas are especially likely to cause metastasis to the ribs and spinal column Cancer of the breast in women and cancer of the prostate in men are most likely to cause pathologic fracture of the ribs

The symptoms of metastatic tumors of the ribs are dull aching pain and most frequently intercostal neuralgia When the lesion is in the upper ribs the pain often has a brachial plexus distribution The patient may discover a swollen or tender region which persists

Often the results of examination are negative if there are no fractures present Examination by physical means is never diagnostic of rib tumors Diagnosis is established by roentgenographic examination When the tumor is si[illegible]

blood from the stomach *hematemesis* The bloody sputum of hemoptysis is bright red contains froth and does not contain food When in doubt about whether there has been hemoptysis or hematemesis ask the patient whether the blood came from the lungs or the stomach The patient can often answer this question correctly and may even state correctly the side of the chest from which the hemoptysis came

When blood is intimately mixed with mucus in the sputum the color varies according to the relative quantities of these two constituents and the length of time after they have been mixed Thus in pneumonia the sputum may have a brick-dust tint (rusty sputum) or it may be outspokenly hemorrhagic Mucohemorrhagic sputum is also met with in hemorrhagic infarction of the lung and in neoplasm

The so-called prune juice sputum (an unnecessary term) is a serohemorrhagic sputum It is met with when a croupous pneumonia is complicated by edema of the lung A peculiar mucohemorrhagic sputum of sticky consistency said to resemble currant jelly (another unnecessary connotation) has been described in those who have neoplasm and sometimes in pulmonary syphilis

Small streaks of bright blood in sputum otherwise of mucous character usually originate in the upper air passages or in the mouth

Blood stained saliva is often brought to the physician by the hysterical and by simpletons It is usually a thin fluid of stale odor and of brownish red color

Color of the Sputum Sputum may be of different colors owing to the admixture of various coloring matters *Black sputum* is seen in coal miners and iron workers *blue sputum* is occasionally seen in workers in dye works *green sputum* is met with in pneumonia with jaundice sometimes in caseous pneumonia and occasionally in pyocyaneus infections *yellow sputum* may be due to hematoidin in pulmonary abscesses or to bile when hepatic abscesses break into the lung In chronic passive congestion of the lung in cardiac disease a *yellowish red* tint may appear owing to the large number of pigment containing cells—heart failure cells—in the sputum

Odor of the Sputum Usually stale the odor of sputum may become putrid from decomposition in the mouth or in the air passages (fetid bronchitis bronchiectasis pulmonary gangrene) There is no odor more sickening than that of fetid sputum

Consistency of the Sputum The consistency of sputum depends on the relative amount of mucus it contains in asthma and sometimes in pneumonia the sputum is so tenacious that it must be wiped from the mouth

Protein Content of the Sputum Sputum arising chiefly from increased secretion of the bronchial mucous membrane as in asthma and in bronchitis is poor in protein whereas sputum arising in inflammations of the lung substance itself for instance or in transudations such as pulmonary edema and chronic passive congestion is rich in protein *The protein loss in the sputum except in hemoptysis* is not of sufficient quantity to be of practical importance The urea content of the sputum closely parallels the concentration of urea in the blood

Particulate Matter in the Sputum Fragments occasionally pieces of pulmonary tissue may be coughed up in gangrene or in pulmonary abscess Pieces of *tumor* and *tumor cells* often appear in the sputum The finding and identification of tumor cells in the sputum are valuable diagnostically and these are searched for in all instances of suspected malignant disease of the lungs or bronchi

Sometimes branched fibrinous casts of the bronchi are coughed up in fibrinous bronchitis in croupous pneumonia or in diphtheria These are recognizable with the unaided eye They are of no particular diagnostic significance When they are small if desired these casts may be isolated by shaking the sputum with water

Curschmann s spirals Charcot Leyden crystals tuberculous lenses and Dittrich s plugs are terms which are the remnants of former days and convey no implications of definite pathologic significance In the decomposing sputum of putrid bronchitis of bronchiectasis and of pulmonary gangrene small yellowish white masses may be present

COUGH A cough consists of a forcible explosive expiration during which the glottis is first closed and then quickly opened The current of air passing between the vocal cords gives rise to a noise that is at first of high pitch becoming lower as the glottis opens

A cough is a defensive mechanism which when properly used helps to cleanse the larynx the trachea and the larger bronchi The sensory stimulus passes up the vagus to the cough center in the medulla oblongata which lies close to the respiratory center

Unnecessary coughing injures the mucous membranes of the air passages *Violent coughing can injure the elasticity of the lung especially in its upper parts* Coughing exerts an important influence on the circulation As results of coughing the intrathoracic pressure is increased the inflow of venous blood is hindered and the outflow of arterial blood is favored so that the arterial pressure may momentarily be increased This sudden heightening of the blood presure may lead *to arterial rupture in atherosclerosis or in aortic aneurysm it accounts for the con* junctival hemorrhages in any patient who has a violent cough Prolonged violent coughing may cause extreme soreness and tenderness in the epigastric muscles

When nothing is expectorated the cough is dry or empty as in cutaneous or in pleural irritation

A hacking *or frequently recurring feeble cough indicates continuous* slight irritation it is met with in acute and chronic catarrh of the upper air passages Violent paroxysms of coughing difficult to allay are common in convalescence from influenza The so called goose cough or brassy cough of aortic aneurysm is characteristic only of a mediastinal tumor with pressure on the recurrent laryngeal nerve The whoop of whooping cough has only to be heard once to be afterward easily recognizable However the whoop is not limited to whooping cough It occurs in any severe irritating cough in either children or adults There is a laryngitis caused by Pfeiffer s organism occurring in adults which often is accompanied *by the whoop* These affections are often termed second attacks of whooping cough

In summary it is well to repeat Pottenger s definition of the mechanisms of cough which consist of (1) a sensory disturbance in the larynx through the superior laryngeal nerves (2) a closure of the glottis through the superior and inferior *laryngeal nerves (3) a contraction of the expiratory muscles* (internal intercostals and abdominal) through the thoracic spinal nerves and (4) relaxation of the inspiratory muscles (diaphragm external intercostals intercartilaginous muscles of the shoulder girdle) through the cervical and thoracic spinal nerves

THE SPUTUM The sputum expectorated on hawking or coughing consists of a mixture of secretions from the mucous membranes with pus blood or other materials given off from the respiratory apparatus The secretions come from the mucous membranes of the larynx trachea and bronchi pharynx and back of the nose *In addition to these secretions the sputum contains saliva* secretions from the mucous membranes of the mouth and food particles

The amount of sputum is extremely variable and depends on the disease process present The largest amounts are met with in certain *bronchial affections* in large bronchiectatic or phthisical cavities in pulmonary edema or when abscesses or empyemas break into the bronchi In such cases the sputum may be brought up in mouthfuls (as much as 1 to 2 liters daily)

Varieties of sputum are designated according to the predominating constituent present *Four main varieties of sputum are described* mucous purulent serous and bloody Other forms of sputum are admixtures of these for example mucopurulent and mucohemorrhagic

Sometimes there is expectoration of pure blood which is *termed hemoptysis* In hemoptysis the blood is coughed up it is distinguished from the vomiting of

whether from primary heart failure or from heart failure secondary to fibrosis of the lungs and the pneumoconioses emphysema or arteriosclerosis

Deficient aeration of the blood such as occurs in tracheal obstruction bronchial asthma uncomplicated emphysema and in the acidosis of uremia or diabetes is an important cause of dyspnea The dyspnea thus produced is enhanced by myocardial insufficiency with an ensuing degree of pulmonary congestion In pulmonary congestion the lungs become stiffened and less distensible and therefore a reflex mechanical factor of dyspnea is added

Dyspnea may also occur as a hysterical manifestation either in the form of *attacks of deep sighing respirations usually following some emotional disturbance* or occasionally even as paroxysms of hyperpnea with deep and labored breathing If the resulting hyperventilation persists long enough alkalosis with tetany ensues

Certain clinical varieties of dyspnea associated with certain phases of respiration are commonly recognized Inspiratory dyspnea is due to obstruction of the larynx or trachea which slows the respiratory rhythm by prolonging and deepening inspira tion it causes apprehension and often anguish to the sufferer and is accompanied by a collapse during inspiration of the soft parts above the sternum and below and between the ribs

Expiratory dyspnea is due either to an advanced loss of pulmonary elasticity as in emphysema or to generalized bronchial spasm as in asthma Dyspnea of effort is a symptom of many types of respiratory disease It is a principal sign of the disturbances of pulmonary circulation caused by cardiovascular diseases

Orthopnea in Pulmonary Disease Orthopnea is dyspnea present even when the patient sits up It represents a serious exhaustión of the respiratory reserve which consists of an urgent need for oxygen and an accumulation of carbon dioxide in the blood Compensation is striven for by an increased depth of respiration this is obtained by prolonging and deepening respiration with little or no increase in rate Orthopnea attends laryngeal and tracheal obstruction the diffuse bronchial spasm of asthma and the diffuse spasmodic obstruction of the earliest stage of bronchiolitis In all conditions attended by an increased rigidity of the lungs for instance in the pneumonias in massive collapse of the lung in all forms of pul monary congestion or edema and in diffuse miliary tuberculosis breathing is shallow and rapid

STRIDOR Air passing in and out of a partially obstructed larynx or trachea may produce a harsh vibrating noise This vibrating noise is termed stridor It is a noise produced in the larger air passages in the same way that rhonchi are pro duced in the smaller ones Stridor may occur during any part of the respiratory cycle It is ordinarily most obvious in inspiration and during coughing imparting a harsh high pitched quality to the cough The obstruction of the larynx and trachea may be intrinsic as for example by a diphtheritic membrane or it may be extrinsic as for example a mediastinal tumor or from kinking of a bronchus such as may be produced by localized fibrosis of the lung The obstruction of a main bronchus is attended by a wheeze not stridor

HOARSENESS In disease inside the thorax the larynx is often secondarily affected either by local inflammation or by paralysis due to pressure on the recurrent laryngeal nerve The dysphonia which ensues may vary from a mild huskiness to aphonia The extent of the loss of voice is not a measure of the seriousness of the lesion

Acute laryngitis causes a dysphonia which passes off as the inflammation re solves The voice may be altered by extrinsic factors also—nasal obstruction tracheal obstruction pharyngitis and chronic disease of the lungs Aphonia or soft whisper *may occur in neurotic patients*

Hoarseness which persists in persons more than 40 years old may be due to a tumor of the larynx either benign or malignant

They consist of particles of tissue or of small masses of pus which have undergone decomposition in the lungs or in the bronchi Sputa containing these materials have a foul fetid odor

Small soft yellowish white particles consisting of fungi are common in pulmonary tuberculosis and in chronic bronchitis In actinomycosis of the lungs these particles are termed sulfur bodies Pulmonary stones may be present in sputum and when the patient inquires about them a correct answer should be given These stones are portions of calcified lymph nodes

CYANOSIS IN PULMONARY DISEASE The blue color of the skin and mucous membranes in cyanosis is due to the presence of sufficient reduced hemoglobin in the capillaries to give the color which shows through the skin A minimal content of 5 gm of reduced hemoglobin per 100 ml of blood is necessary before the blue color is apparent In profound anemia however with a hemoglobin content of less than 5 gm the amount of reduced hemoglobin cannot reach this minimal content even in the presence of marked anoxia and thus the profoundly anemic patient will not be cyanosed The greater the amount of reduced hemoglobin the deeper the cyanosis It is best seen in the lips nose cheeks ears fingers and toes It can be temporarily abolished by pressing out the blood from the part The intensity of the color depends also on whether the small peripheral vessels are dilated or contracted

The blood in the capillary vessels may contain an excess of reduced hemoglobin either because it left the heart in this condition or because there is an abnormal excessive change of oxyhemoglobin to reduced hemoglobin while the blood is passing through the peripheral capillary system

Abnormally slow passage of the blood through the peripheral capillaries allows the tissues to take more oxygen from it than usual and so to produce more reduced hemoglobin Slowing of the peripheral circulation is a normal response to cold and mild degrees of local blueness are compatible with good health When capillary stasis or poor blood flow is associated either with general circulatory failure or with some form of local vascular paralysis it is indicative of serious disease

It is not always easy to determine the cause of cyanosis in a particular patient whether it is respiratory or circulatory or a combination of both When the capillaries are engorged the pulse full the flow rapid there is in addition to the deficient oxygenation in the lungs an associated carbon dioxide retention with peripheral vasodilatation and an inadequate circulation In some instances of inadequate circulation the cyanosis is deep and florid and the patient breathes heavily while in others the skin may be pale cold and wet with leaden livid cyanosis indicating poorly filled capillaries in a patient who has respiratory and circulatory failure a rapid feeble pulse and quick shallow breathing

Cyanosis may also result from the formation in the blood of certain derivatives of hemoglobin Methemoglobin is produced by large doses of drugs such as chlorates coal tar derivatives and sulfonamides These drugs may also determine the formation of sulfmethemoglobin which gives rise to a leaden type of cyanosis

DYSPNEA IN PULMONARY DISEASE Dyspnea is characterized by difficult painful or disordered breathing Hyperpnea is increased breathing A healthy person is not conscious of any increase in respiration until the normal rate is doubled

The obvious causes of dyspnea are many for instance the various diseases of the lungs and pleura cardiac failure anemia the acidosis of uremia or diabetes increased intracranial pressure and hysteria The important causes are chemical alterations in the blood flowing through the respiratory center and local conditions in the lungs which make them more rigid and less easily distensible and cause rapid and shallow breathing Rigidity or decreased distensibility of the lungs is by far the more usual factor in the dyspnea It determines the dyspnea of pneumonia of pulmonary edema and of massive collapse of the lungs Pulmonary congestion with consequent rigidity of the pulmonary tissue is the cause of cardiac dyspnea

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EXAMINATION OF THE LUNGS AND PLEURAE

In studying clinically the condition of the lungs and pleurae the physician employs the aids of inspection palpation mensuration percussion auscultation and exploratory puncture The roentgenologic examination of the lungs and pleurae often aided by the intrabronchial use of radiopaque substances is more precise and reliable than any other examination by physical means The bronchoscopic examination is more precise for the purposes of diagnosis in certain instances than any other examination

The Respiratory Movements The lungs make no active movements of their own during respiration but simply follow passively the movements of the wall of the thorax and of the diaphragm Two main types of inspiratory expansion of the thorax are seen In the *costo abdominal* type the expansion is due chiefly to the descent of the diaphragm and less to the elevation of the ribs In the *costal* type the opposite is the case the expansion being due chiefly to elevation of the ribs

The diminution in size of the thorax during expiration depends normally on the elasticity of the lungs and ribs not on muscular contraction

The times occupied by inspiration and expiration respectively are approximately equal and there is no pause between the two

FREQUENCY AND RHYTHM OF THE RESPIRATIONS The number of breaths taken by *healthy adults varies between 16 and 20 per minute The rate in disease* may be increased or decreased

Increase of the frequency of respiration (dyspnea or tachypnea) is met with in most diseases of the respiratory system in many diseases of the heart and in those diseases of the abdomen that hinder the movements of the diaphragm In dyspnea the number of respirations may reach 40 60 or more per minute In certain diseases of the nervous system excessive tachypnea may occur in paroxysms

Mechanisms of the production of tachypnea are discussed under Heart Disease (Chapter 10)

Slowing of the respiration (bradypnea) may be met with in affections of the brain or of its membranes in irritations of the respiratory center in *the medulla in* severe infections and in uremia

Cheyne Stokes Breathing In this form of respiration periods of complete cessation of the respiratory movements alternate with periods in which the respirations slowly increase in frequency and volume to a maximum and then again decrease until they cease

The phenomenon is met with in many normal persons while asleep and therefore is inconsequential in illness despite the fact that it is observed in an exaggerated form in many patients who are seriously or critically ill During the apneic phase these very ill patients are sleepy and the pupils are contracted and nonreactive as the respiratory movements return the psyche awakens and the pupils *dilate and* again become responsive

In *Biot s breathing* the respiratory pauses are long (from 5 to 30 seconds or more) in periods which may recur more or less regularly or wholly irregularly

Dyspnea is observed normally on exercise and in disease either of the lungs or of the circulatory apparatus As dyspnea increases the patient sits up in bed with his head arms and thorax held rigid panting for breath and the condition is then termed orthopnea

DETERMINATION OF THE VOLUME OF THE INSPIRED AIR AND OF THE EXPIRED AIR (SPIROMETRY) The measurement of the air entering and leaving *the lungs* in each phase of respiration can be made by means of a spirometer

Certain terms which have been established in spirometry should be understood since these terms may be employed in description of minute output of *the heart* determined by gasometric methods (carbon monoxide) and occasionally on patients

who have heart disease By *vital capacity* is meant the amount of air that after the deepest possible inspiration can be expelled on full expiration in healthy men it varies from 3 000 to 4 000 ml in women it varies between 2 000 and 3 000 ml The vital capacity is smaller in old age as well as in all diseases of the respiratory system

Tests of pulmonary function offer a method of appraising pulmonary disabilities objectively They are of value in determining the absence of pathologic alterations in some of the nonpulmonary types of dyspnea as seen in neurocirculatory asthenia and hysteria the course of various pulmonary diseases can be followed objectively and the preoperative use of the tests will indicate fairly accurately the results of such surgical procedures as collapse therapy lobectomy and pneumonectomy

The increased interest in pulmonary physiology since surgical operations on the lungs and the heart have become commoner has brought about a revision of the terminology used to describe the various components of respiration

Inspiratory Reserve (Complemental Air) This measures the maximal amount of air which can be drawn into the lungs using the resting expiratory level as the starting point Normally this amounts to 3 000 ml

Expiratory Reserve (Supplemental Air) This measures the maximal amount of air which can be expired using the resting expiratory level as the starting point Normally this amounts to 1 000 ml

Residual Air (Residual Capacity) This represents the air remaining in the lungs following a maximal expiratory effort This quantity cannot be measured spirographically and its determination depends on measuring the amount of nitrogen displaced by breath ing 100 per cent oxygen

Functional Residual Capacity This represents the quantity of air remaining in the lungs after the completion of a quiet respiration It is the sum of the residual air and the expiratory reserve

Maximal Breathing Capacity This is a measure of the maximal volume of air that can be breathed in a unit of time The maximal breathing capacity differs from the vital capacity in that it expresses a time relationship The same information can be obtained by measuring the vital capacity and relating it to the time required for expiration

The amount of air taken in during a minute of quiet respiration varies from 5 to 7 liters

The vital capacity of the lungs enlarges on bodily exercise and in pathologic states accompanied by dyspnea

INTERCOSTAL RETRACTION AND EXPANSION On inspection of the thorax it is well to observe the rate of the respirations and record it The progression of expan sion of the lungs is followed during inspiration with particular reference to the lack of filling and consequent expansion in any part of the lung and especially at the apices In the adult in a normal state of nutrition there is no noticeable expansion or retraction in the intercostal spaces

The Diaphragmatic Phenomenon (Litten s Sign) In emaciated adults the res piratory movements of the lower margin of the lungs can sometimes be observed as a furrow descending during inspiration over the intercostal spaces especially in the lower part of the right side of the thorax (Litten s sign) This is due to the separation of the diaphragm from the thoracic wall when it contracts before the lungs can follow it the atmospheric pressure causes a depression of the soft parts

Litten s sign disappears in inflammatory diseases of the lungs pleura and pleural adhesions to reappear in convalescence if inflammatory disease heals without scar formation

Mensuration of the Thorax By passing a tape around the thorax just above the nipples the patient s arms being held out from the sides the circumference of the thorax is measured On physical examinations for entrance to the army or navy and for life insurance this value is used as a measure of the development of

the thorax and serves as a clue to the general constitution of the person In healthy young men the circumference should exceed 32 inches (about 81 cm)

More important as regards the state of the lungs themselves *is the difference in circumference on deep inspiration and on deep expiration* Normally this difference amounts to about 2⅘ inches (7 cm) or more

The comparison of the *circumference of each of the two halves* of the thorax shows normally a difference of from 0 5 cm to 2 5 cm in favor of the right side In an asymmetric thorax owing to unilateral expansion or retraction slight differences in circumference of the two halves of the chest can be easily recognized

Exact records of the form of the thorax are made with a *cyrtometer*

Palpation of the Thorax Over the Lungs and Pleurae A part of the palpation of the thorax is the testing of the vocal fremitus the vibration of the voice propagated to the thoracic wall through the bronchi and pulmonary tissue The test *is made by placing the two hands on symmetric portions of the thoracic wall and asking the patient to repeat in a deep voice 1 2 3 or 99 In health the vocal* fremitus perhaps is more marked on the right than on the left side In women the vocal fremitus is weaker than in men Vocal fremitus sometimes is helpful in deciding whether dullness over the lower part of the thorax is due to infiltration of the lung or to pleural effusion increased vocal fremitus indicates better conduction from the larynx to the lung surface (infiltration) while enfeeblement or abolition of the fremitus suggests pleural effusion or pneumothorax However in massive pneumonia with plugging of the bronchus the propagation of the voice may be temporarily interfered with again in pleural effusion there may be less diminution of the vocal fremitus than might be expected because pleural adhesions uniting the *lung to the wall of the thorax may favor the conduction of the voice* tremor or an atelectasis due to the pressure of the exudate may increase the vocal fremitus more than the effusion weakens it

Percussion Over the Lungs and Pleurae In percussion a shock is given by the percussion stroke which gives rise to sounds that vary with the elasticity (capacity for vibration) of the parts struck the greater the elasticity the more marked is the production of sound

PRINCIPLES OF PERCUSSION Airless bodies enter into sound producing vibration only when they are rigid or in a certain state of tension In the human body bones alone correspond to such a state

The sounds yielded by soft parts when percussed depend on the air or gas content *If the air or gas is contained in large cavities* it is set into vibration and along with it the wall of the cavity is started vibrating if the air is distributed and confined throughout the whole tissue as in the lungs the air and the tissue vibrate together on percussion Percussion then permits of an approximate delineation of the limits of juxtaposed airless and air containing structures

DIRECT PERCUSSION The fingers are arranged in a plane with the middle finger stiffened and slightly flexed toward the palm and *to produce the sounds* deliver the

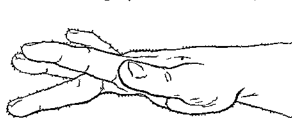


Fig 9 4 Direct percussion employing the stiffened *middle finger*



Fig 9 5 Direct percussion with the end of the middle *finger*

stroke directly on the surface of the thorax with the middle finger without any intermediary plessimeter (Fig 9-4) A better method is to use the end of the finger as in Figure 9 5

INDIRECT PERCUSSION In this method the blow instead of being struck directly on the body surface is delivered on some intermediate body placed upon the part under study This intermediate body is called a plessimeter or pleximeter As a *plessimeter* the middle finger called a plessimeter finger is used (Fig 9 6) To ascertain the condition of the structures behind the clavicle the clavicle itself often is used as a plessimeter

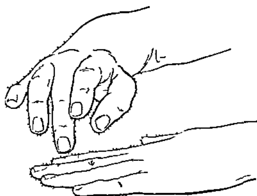


Fig 9 6 Indirect percussion employing the middle finger as a plessimeter

The plessimeter finger is most often used because it permits judgment of the feeling of resistance thus yielding some information accessory to the percussion sounds It is easy to approximate the finger to the surface such as the intercostal spaces and supraclavicular fossae

As a *percussor* or *plessor* it is customary to use the middle finger of the right hand flexed to an obtuse angle at the two distal terminal joints and slightly flexed at the proximal joint of the finger (Fig 9 6) The stroke is delivered entirely from the wrist the forearm remaining in position With practice one soon learns how best to deliver the stroke As a rule the percussing finger should be allowed to rebound immediately from the plessimeter after it has struck in order that the plessimeter may freely vibrate with the part beneath In gentle percussion the plessimeter finger should lie in smooth contact with the part under study for deep percussion it may be firmly pressed against this part

In *orthopercussion* the force of the blow is directed exactly perpendicular to the surface

The *strength of the percussion stroke* should vary greatly according to the conditions of the examination When the soft parts over the organ under examination are thick the stroke must of course be harder to set that organ in vibration

In the determination of so called *deep or relative dullness* of the heart however it is necessary to use a somewhat stronger stroke though even here ones does best to use percussion of medium force rather than the very strong percussion so often employed

THE INTENSITY AND QUALITY OF THE SOUNDS PRODUCED ON PERCUSSION In percussion of the thorax as elsewhere attention is directed regarding the sounds produced to their *loudness* their *pitch* and their *clang* or *timbre*

Comparative Percussion of the Lungs In health the pulmonary resonance on percussion varies somewhat over different regions of the thorax corresponding to the variable thickness of the soft parts and to the variable volume of lung beneath In some places the sound is louder and fuller than in others By repeated percussion of the healthy chests of different persons there gradually become fixed in mind memories of the normal sounds heard and of the normal resistance felt on percussion over the lungs It is learned how much muffling or dulling of the sound may be due to varying degrees of obesity and to different degrees of muscular development The percussion sound is always clearer in front and at the sides than behind In fat persons the resonance on percussion over the back may be impossible to evaluate

It is always well to compare the percussion sounds yielded by symmetric parts

on the two sides of the chest, beginning with percussion of the supraclavicular fossae above and passing downward on the two sides over the several intercostal spaces making sure to percuss always with the same force and to direct the stroke in the same way

The breathing should be quiet and normal during percussion The loudest sounds are normally met with between the third and the fifth ribs in front

It is wasted time to percuss over the tympanitic space of Traube (fourth to sixth rib on) owing to the presence of the stomach under the left side and front of the lower part of the thorax In comparative percussion of the two sides of the chest asymmetries of the two sides of the thoracic wall are important A slight grade of scoliosis may affect the sounds

The loudness of the percussion sounds depends essentially on the amount of air or gas in the organ examined

The terms clear and dull refer to the *intensity* of the sound Clear and dull are used *clinically in a different sense from that of ordinary speech Clinically reference to the* loudness of sound is to the amplitude of the sound waves that strike the ear drum This intensity depends on the capacity for vibration and the strength of the percussion blow In comparative percussion therefore use exactly the same force and avoid confirming a preconceived opinion by unequal *force of the percussion blows* for unequal force will produce unequal sounds and dullness may thus be simulated

The *pitch* of a tone depends on the number of vibrations per second The greater the number of vibrations the higher the tone Percussion sounds are always *noises* composed of a large number of individual *tones* The lowest tones are met with over the distended lungs in pulmonary emphysema A percussion sound therefore that contains very low tones is usually loud clear and full

The ear should be carefully exercised in the appreciation of the lower tones on percussion When one tone is dominant as in tympanitic sounds the height of its pitch is easily recognizable to one who has a good tone pitch sense The recognition of differences in the pitch of the dominant tone in tympanitic sounds lies at the basis of the recognition of alterations in pitch

In general it may be said that a *tympanitic sound* over a lung except in Traube s space which is situated in the lower part of the left lung indicates an abnormal condition within the thorax such as pulmonary cavities bronchiectatic cavities abdominal viscera dislocated into the thorax or a change in the structure of the lung that leads to loss of the tension of the alveolar walls for example atelectasis of the lung from bronchial occlusion or from compression due to high position of the diaphragm large pericardial effusions or pleural effusions or in edema of the lung in certain stages of pneumonia when the alveolar septa have changed in texture from the inflammation though the air content is not yet essentially diminished or in bronchial pneumonia in which the bronchi in airless tissue may give rise to a tympanitic *note*

A *metallic* or *amphoric sound* on percussion depends on the presence of high overtones accompanying and obscuring a low fundamental tone It arises in large gas containing cavities with tense walls It will appear in the blown up stomach if the distention is sufficiently increased To grow familiar with this sound it may be elicited by thumping over the cheek after the mouth has been so strongly distended by air that the tympanitic sound present on less distention disappears

A *cracked pot sound* (or metallic clink) arises on strong percussion if air or gas is thereby driven out of a cavity through a narrow opening

THE FEELING OF RESISTANCE ON PERCUSSION It may be possible to draw more precise conclusions from percussion if the examiner not only listens but feels with the finger which is being percussed The sense of resistance *felt by the fingers* on percussion varies with the compressibility of the part percussed It is readily admitted that percussion areas do not correspond to anatomic projections but by practice of finding these areas in the normal thorax pathologic changes can often be recognized and the clinician is better for having this

The Upper Limits of the Pulmonary Resonance on Percussion Normally the upper limits are in the same position on the two sides On each side the limit extends for a distance of from 1 1/5 to 1 3/5 inches (about 3 to 4 cm) above the clavicle at the level of the spine of the sixth or seventh cervical vertebra and behind goes over the edge of the trapezius muscle obliquely downward and mediad into the posterior medial pulmonary limit at the level of the spine of the second thoracic vertebra (Kronig s fields of pulmonary resonance at the apices)

Normally the upper margin of the lung reaches a point above the clavicle cor responding to the position of the tubercle on the first rib which can be felt on palpation between the heads of the sternocleidomastoid muscle

The Lower Limits of the Pulmonary Resonance on Percussion These can be approximately determined by percussion

The lower limit of the pulmonary resonance varies with the phases of respiration and with the position of the patient On forced inspiration the boundaries may descend 4/5 to 1 1/5 inches (about 2 to 3 cm) below the level found in quiet breath ing In the dorsal decubitus the lower margin is [illegible]/5 to 4/5 inch (about 1 to 2 cm) below its position in the sitting posture in the lateral decubitus that of the upper lung may descend 1 1/5 to 1 3/5 inches (about 3 to 4 cm) lower

Abnormal Position of the Boundaries of the Lungs The determination of the pulmonary limits may in pathologic states show a general expansion a displace ment only of the lower boundaries a general contraction or a displacement of either the lower or the upper limit

General expansion of the pulmonary limits is met with in chronic emphysema and in the temporary inflation of the lungs due to tracheal or laryngeal stenosis diffuse bronchitis and bronchiolitis or asthma The lower limit may be lower than normal from compensatory emphysema of the lower part of the lung when the upper part is diseased for example in apical tuberculosis or when the opposite lung is diseased

General contraction of the pulmonary limits is met with when the total amount of air-containing tissue in the lungs is diminished from any cause that is prevention of entrance of air into the lung or compression of the lung from thoracic tumors or abdominal distention

The *lower limit of a lung may be higher* than normal when the diaphragm is high as in abdominal distention or in retraction of a lung or when parts of the lung are deprived of air as in bronchostenosis A very common cause for displacement of the lower limit of pulmonary resonance upward is the accumulation of fluid in the pleural cavities When the pleura is nonadherent the fluid tends to accumulate chiefly in the lateral parts the line of the dullness does not as a rule run horizontally but in a curve the highest point of which lies approximately in the posterior axillary line descending rather abruptly in front and more gradually behind (S shaped line of Gardner)

Retraction of the upper limit is most often due to pulmonary tuberculosis The retraction of the apex is usually first demonstrable at the upper medial margin in front

DULLNESS AND FLATNESS ON PERCUSSION The percussion sound is dull in all conditions in which the air content of the lung percussed is less than normal

When the lung is separated from the thoracic wall by fluid there must be at least 500 ml to yield dullness on percussion An immediate change of the dullness with change of position indicates the simultaneous presence of air and fluid

Auscultation of the Lungs On auscultation of the lungs the examiner listens (with the aid of a stethoscope) to the sounds that accompany inspiration and expira tion and the voice sounds that are audible over the thorax Application of the ear to the thoracic wall is no longer employed unless the stethoscope has been left at home Under these circumstances a great deal of information may be gained

The voice sounds are better heard with the naked ear than with the aid of a stethoscope but the breath sounds and especially some of the modifications that these undergo in disease as well as some of the sounds that accompany them are better recognized and localized by the use of the stethoscope

From Current Comment in the *Journal of the American Medical Association* (Dec 24 1949) on the auscultatory respiratory murmur the following comment is quoted

> A new appraisal of auscultation which is based on clinical experience and facts in physics and physiology discovered since Laennec s time is claimed to show that the respiratory murmur is composed of sound vibrations originating in all component parts of the respiratory mechanism It is now maintained that the respiratory murmur cannot be caused by air rushing through the larynx and impinging on the bronchial walls and dilating the air cells because the lungs always contain residual air which stops the force of the incoming current and causes the air to enter the finer bronchi and air cells by diffusion The sound vibrations causing the vesicular murmur are said to originate in the respiratory mechanism in the anterior lateral and lower portions of the thorax where the air cells predominate the musculature is light and the bony cage is elastic That part of the respiratory mechanism which normally causes the bronchial murmur is found in the superior portion of the thorax and is most distinct posteriorly Here the sound vibrations are produced by the air current the larger bronchi a comparatively small amount of pulmonary tissue and air cells a heavy musculature and the least elastic portion of the bony cage

THE BREATH SOUNDS The breath sounds present over the alveolar masses of the lung are called *vesicular breathing* The breath sounds present over the larger air tubes during respiration are termed *bronchial breathing*

In disease states when there is secretion of excessive mucus swelling of the bronchial mucous membrane or fibrinous exudation on the surfaces of the pleurae certain accessory or *adventitious sounds* arise These sounds are termed rales crepitation friction sounds

The breath sounds normally are loudest just beneath the clavicles The sounds are feebler over the apices

MODIFICATIONS OF VESICULAR BREATHING Accentuated vesicular breathing is heard normally in children and hence is often spoken of as puerile breathing Puerile breathing may be audible until puberty more often in girls than in boys

Accentuated breathing is heard in the neighborhood of infiltrated regions especially *at the apex in incipient pulmonary tuberculosis* and in a healthy lung close to a consolidating region at the beginning of pneumonia

Sometimes there is *prolongation of the expiratory sound* of vesicular breathing so that this becomes as long as or even longer than the inspiratory sound A prolongation of the expiratory part of the vesicular breath sounds often indicates an expiratory dyspnea such as emphysema asthma or bronchitis If the distribution is limited to the base of the right lung it may indicate the beginning of congestive heart failure

If the inspiratory sound instead of being continuous is interrupted so as to consist of two or more parts the inspiration assumes a jerking or saccadic character it is *cog wheel breathing* Cog wheel breathing is often met with in persons of feeble musculature in nervous people and in children after crying

BRONCHIAL BREATHING Bronchial breathing is audible in healthy persons over the spinous process of the seventh cervical vertebra corresponding to the position of the bifurcation of the trachea and thence downward to the spine of the *fifth* thoracic vertebra

The loudness of bronchial breathing is not important It is the aspirating hollow or reverberating character of the sound that is peculiar and it is often well marked when the sounds are weak The expiratory portion of the sound in tubular breathing

may and usually does approach the inspiratory sound in intensity its duration is great In some instances however expiration is inaudible in tubular breathing

Pathologic Bronchial Breathing The solidifications of the lung over which pathologic bronchial breathing becomes audible include (1) infiltrations due to pneumonia tuberculosis or hemorrhagic infarction (2) compression behind pleural exudates and tumors and (3) atelectasis The solid regions under ideal conditions which are seldom present must be either near the surface or be connected with it by means of a good sound conducting medium moreover the communicating bronchi must be open Cavitation resulting from the destruction of air sacs over which pathologic bronchial breathing may become audible includes bronchiectatic cavities tuberculous cavities and larger emphysematous cavities The communicating bronchi must be open and the cavities must be near the surface or must communicate with it by sound conducting media

Mixed Breathing or Bronchovesicular Breathing The term mixed breathing or bronchovesicular breathing is used too frequently and perhaps in some instances for the purpose of mental hedging

Over normal lungs or pleuritic exudates or when loud rales are present the respiratory murmur may be so feeble that the character of the breath sounds cannot be recognized Bronchovesicular breathing simply means the presence of both bronchial and vesicular elements in the breath sounds

Origin of the Voice Sounds In auscultation of the voice sounds two processes are important (1) the production of fundamental tones in the larynx where the vocal cords of the glottis act like a reed instrument and (2) the process of articulation in which the different tones produced in the larynx are modified by changes in shape of the oral cavity and adjacent cavities the larynx itself being unable to articulate The oral cavity in association with the laryngeal and nasal cavities constitutes a resonating chamber As such it intensifies that particular tone or overtone of the sound produced in the larynx the wavelength of which corresponds in resonance to the dimensions of the cavity Variations in the shape of the cavity will modify its function as a resonating chamber in that it will vibrate in unison with notes of different wavelengths or pitch Thus through changes in the resonating chamber different overtones may be intensified or weakened and corresponding changes in the voice sounds produced In whispering there is articulation of the breath sounds only the glottis being passive and giving rise to no loud tones

When auscultation is made over the healthy lungs of a person pronouncing the number 99 in a voice of low pitch the audible voice sounds are nearly everywhere weak and muffled instead of being loud and clear like those audible over the larynx When loud clear voice sounds are audible directly over the bronchi of the healthy thorax the condition is known as bronchophony The term bronchophony includes all such sounds audible in health and all degrees of clearness greater than this in disease

If it is observed that the sounds have become less clear than formerly especially if they suddenly disappear over an area occlusion of the bronchi is often the cause of the disappearance of the sounds

Increased clearness or distinctness of the voice sounds is termed pathologic bronchophony This term is applied when sounds as clear as or clearer than those heard over the upper thoracic part of the spinal column in health are met with in any other part of the thorax The phenomenon is due either to increased conducting power or to increased reflecting power of the lung tissue The conducting power of the spongy structure of the lung is increased by any change that makes the pulmonary structure more homogeneous

Whispered bronchophony is sometimes clearer than that of the spoken voice The terms pectoriloquy and whispered pectoriloquy are sometimes applied when the sounds are very clear apparently close to the ear and exactly circumscribed

A special kind of definite bronchophony known as egophony is sometimes heard above the level of the fluid in large pleural effusions that cause partial compression

and narrowing of the bronchi The voice sounds have a definite nasal quality which is rhythmically intensified and interrupted

Over large cavities containing air, and especially over a pneumothorax, the voice sounds may have an amphoric metallic ringing quality

Auscultation of the voice sounds yields information comparable in large part to that afforded by palpation of the vocal fremitus when the voice is not strong enough or deep enough to yield a palpable fremitus auscultation of the sounds may give information not otherwise obtainable It is well to learn how to correlate and do these two examinations synchronously

ACCESSORY OR ADVENTITIOUS RESPIRATORY SOUNDS In addition to the natural breath and voice sounds and the changes these undergo in disease certain sounds occur inside and outside the lung wholly additional to these Of such adventitious respiratory sounds two great groups are distinguished (1) rales or crackles and rhonchi and (2) friction sounds All rales are caused by moisture in the lungs but they are classified as (1) rhonchi and (2) rales

Rhonchi and Rales These sounds arise during respiration from interference with movements through the air passages of the air breathed These interferences with the movements of the breathed air result from the presence of fluid mucus serum blood or water in bronchi and in cavities leading often to the formation of air bubbles which may interfere with the movement of different masses of mucus or secretion and thus add noise The sudden separation of sticky mucous membranes or the passage of air through abnormal local narrowings produces sounds

These sounds may be loud or feeble numerous or few they are often inspiratory in time though they may be audible during expiration On testing for the presence of rales the patient is asked to take a deep breath and end it with a short cough It is important to notice whether the rales disappear or are modified on coughing

Rhonchi are long whistling or snoring sounds that die away slowly They are due to the vibrations of tough mucus attached to the walls of the bronchi or to the passage of the air through lumina narrowed by spasm of the bronchioles or by swelling of their mucous membrane They are often accompanied by palpable vibration of the thoracic wall The sounds when low pitched have a snoring character and are termed sonorous rhonchi If these sounds are high pitched they are more whistling or piping in character and are termed sibilant rhonchi

Rales arise when fluid is present in the air passages They are crackling or *bubbling sounds In contrast with the more continuous* chiefly expiratory sibilant and sonorous rhonchi rales are interrupted bubbling or crackling sounds heard during inspiration

The *moist rales* are sometimes classified according to the apparent size of the bubbles or crackles the largest bubbles being known as gurgling rales those next in size as medium rales and those of smallest size as crepitant rales The sound of the so called crepitant rale can be very well imitated by *rubbing a lock* of the hair between the fingers close to the ear

The *crepitant rale* no matter in which condition it is met with is believed to be due to the opening up of collapsed air sacs or minute bronchioles It is distinguishable from other *moist rales chiefly by the short duration or small size* of each of the successive crepitations and by the large number of crepitations attending each inspiration

Ringing or consonating rales are bubbling sounds arising in a cavity or in a larger bronchus and well propagated to the surface through infiltrated pulmonary tissue the higher tones are strengthened through resonance in the bronchi This ringing character of a rale is therefore of diagnostic importance in infiltrations of the lung Consonating rales occur under the same conditions in general as does bronchial breathing

Metallic ringing rales occur under conditions similar to those in which a metallic clang on percussion and metallic bronchial breathing occur The so-called *metallic tinkling* is a single sound heard intermittently by the physician or the patient It occurs most often in pyopneumothorax or in large tuberculous cavities

PLEURAL FRICTION SOUNDS Pleural friction sounds arise during respiration from the rubbing together of the pleural surfaces roughened by some pathologic process such as fibrinous or inflammatory exudates

These sounds are heard most frequently when the pathologic process is situated between the costal pleura and the pulmonary pleura between the pulmonary pleura and the mediastinal pleura or between the pulmonary and the diaphragmatic pleura The sounds may be fine coarse or scratching or creaking They may be intense or feeble

The softer gentler friction sounds may sound like vesicular breathing the coarser crackling friction sounds may sound like rales The pleural friction sounds however are usually accompanied by pain they sound as if they were close to the ear and they are intensified by pressure of the stethoscope They are often accompanied by a palpable friction fremitus They do not disappear nor are they modified on coughing Pleural friction sounds indicate the presence of a dry pleurisy in the region in which they are audible Friction sounds sometimes become audible in tumors of the pleura or when the pleura is abnormally dry as in severe dehydration

Near the heart pleural friction may be heard which is synchronous with the heart s action owing to the rubbing of the pericardial pleura against the adjacent pulmonary pleura This is termed pleuropericardial friction

SUCCUSSION SPLASH COIN SOUND When the pleural cavity contains both liquid and air and the patient is given a vigorous shaking while the physician listens to the thorax a splashing sound is heard This is termed the succussion splash The patient may be able to hear it and to produce it at will and may complain of it Even if this splash has never been heard by the physician he should know about it in order to interpret it if the patient complains of it This splash is different from the one similarly produced in the stomach

A succussion splash is usually due to pneumothorax it is occasionally heard in the absence of pneumothorax when large cavities exist in the lung

The coin sound is hardly a diagnostic test It is elicited by percussing with coins on the back of the thorax at the level of a cavity and by listening with a stethoscope in the front of the thorax at the same level

Roentgenographic Examination of the Thorax The *efficiency and limitations* of roentgenographic examination of the chest in the diagnosis of thoracic diseases by this means have been summarized by Rigler essentially as follows

1 Pulmonary edema can be detected before the onset of appreciable symptoms or within a few hours thereafter It is detectable in the roentgenogram from 6 to 12 hours before the appearance of physical signs

2 Pleural effusion in amounts of 100 ml or more can be detected if proper technic is used This amount is compared with 500 ml of fluid necessary for demonstration by physical examination

3 Bacterial pneumonias give roentgenographic evidences of their presence in as short a time as 2 hours most frequently within 6 hours and almost invariably within 12 hours of the onset of definite symptoms

4 Atypical or virus pneumonias may give no distinctive roentgenographic findings for 24 to 48 hours after the onset of symptoms

5 Pulmonary tuberculosis of the ordinary chronic type is not demonstrable until 8 to 20 weeks after the exposure

6 Acute miliary tuberculosis invariably produces symptoms before the development of roentgenographic signs The latter may not be present for as long as 7 weeks after the onset of symptoms

7 Nodular lesions such as those produced by metastasis or nodular tuberculosis are demonstrable when their diameter is 3 mm or larger Even lesions of this size will

not be seen if multiple films in various positions are not made or if the lesions are unfavorably located

8 Metastatic lesions of miliary size such as those produced by miliary tuberculosis may not be visible until numerous or large in size

9 All metastatic lesions are roentgenographically visible before the onset of symptoms or physical signs

10 Bronchogenic carcinoma almost invariably gives positive roentgenographic signs when symptoms are present if thorough examination is done There are a few exceptions to this rule In most instances roentgenographic evidences of abnormality will be present before the onset of any respiratory symptoms due to bronchogenic carcinoma Complete examination is necessary however to establish these evidences

Reliability of interpretation of roentgenographic evidence of diseases of the lungs and pleurae requires training skill and experience The clinical management and even the details of treatment in thoracic disease often are based on the interpretation of shadows observed in roentgenograms Physicians may hesitate to question the reliability of this method of examination although they realize well the fallibility of other diagnostic procedures that appear less mysterious to them Shadows that appear on films must be interpreted in pathologic terms

An editorial in the *Journal of the American Medical Association* of December 29 1951 discussed a project designed to record different opinions in regard to interpretation of roentgenograms of the chest The project involved the recording of opinions by six experienced observers three of whom were radiologists and three of whom were internists regarding the stability of lesions of pulmonary tuberculosis as recorded on single roentgenograms (postero anterior projections) taken at intervals of three months Each observer independently reported his opinion of each pair of films and subsequently re examined the same series of films without reference to his original interpretation The data thus recorded were analyzed by competent biostatisticians who were able to measure the inconsistencies of interpretation and relate the scores of each reader with the others and with his own previous opinions It was revealed that a second observer will disagree with the first observer in about one third of all instances and what is more surprising when the same observer reviews the same film a second time after an appropriately long interval he will find disagreement between his two opinions in about one fifth of cases studied

These results in no way decrease the recognized necessity and value of roentgenographic examination of the chest but they do emphasize the fact long known to many physicians experienced in thoracic disease that clinical management of thoracic disease must be based on many types of clinical and laboratory data and not on roentgenography alone Furthermore the single postero anterior chest film frequently does not constitute complete roentgenologic examination for fluoroscopy stereoscopy tomography and specially projected views may add materially to the accuracy of the examination in special circumstances However with all of this information available it must be realized that there is no substitute for the clinician whose difficult task involves synthesis of different types of evidence in formulating a program of management for the patient In dealing with thoracic disease the physician relies heavily on roentgenography but realizes that the opinion of the dynamic status of pulmonary disease would in some instances be reversed if a second expert viewed the films or even if the same observer should submit a second opinion after he has forgotten the first report

COMMENT

On examining the thorax by the physical methods heretofore described it is appropriate to try to draw inferences regarding the air content of the underlying lungs and the physical state of the pleural cavities Subsequently it is wise to seek the pathogenetic explanation of these physical conditions of the lungs and pleurae The methods of physical examination by employment of the special senses of the

physician are now known to be only approximately accurate but physicians when trained are grateful for the information they may give

DISEASES OF THE PLEURA

Pleural Pain The *parietal pleura* is innervated by sensory nerve fibers from the intercostal nerves The ability to locate sensory impressions by the pleura is acute everywhere but seems more highly developed in the anterior and lateral aspects

The *peripheral regions of the diaphragmatic pleura* derive their sensory nerves from the phrenic nerve and the last six intercostals The intercostals supply a peripheral rim of the diaphragmatic pleura which is 1 to 3 inches (2 5 to 7 6 cm) wide anteriorly and a segment corresponding to the posterior third of the membrane Irritation of the areas gives rise to pain in the lower part of the thorax on the same side and in the lumbar region or in the abdomen The *central portion* is innervated by the phrenic nerve Irritation of this portion sets up pain in the neck

The afferent impulses reach the cervical cord through the phrenic nerve trunk and pass thence to the brain where the sensorium registers pain in the somatic segment corresponding to the entrance of the phrenic nerve thus the pain is a re ferred pain

Disease or stimulation of the *peripheral or posterior portion of the diaphragm* produces pain which spreads over the lower part of the thorax and epigastrium at times extending down over all of the abdomen on the same side

Capps has observed that the distribution of the pain depends on the degree of irritation and its prolongation The more intense the irritation the greater is the tendency of the pain to spread down over the lower portion of the abdomen and in an occasional instance it may extend over both sides of the abdomen or to the lumbar region The pain may be associated with detectable hyperalgesia of the skin and superficial tissues on pressure

In some individuals who have diaphragmatic pleurisy the pain is referred to the neck along the area of the trapezius ridge The neck pain is spontaneous or produced by deep breathing or cough and is well localized by the patient with the tip of the finger Pressure over this point is painful

PARIETAL PLEURISY The pain in pleurisy confined to the *upper lobes of the lungs* is always situated in the upper scapular and shoulder region In *pleurisy over the lower lobes* the pain is commonly restricted to the lateral and anterior aspect of the lower part of the thorax

Localized pleurisy usually produces a sharp stitch like pain on cough or deep inspiration directly over the region of the pleura involved provided there is mobility between the parietal and visceral pleurae Pleurisy without pain even though a friction rub is present may occur

VISCERAL PLEURA The visceral pleura is insensitive

Pleurisy for the sake of description has often been referred to as dry pleurisy and wet pleurisy or pleurisy with effusion The nature of the effusion is designated by appropriate combinations of word roots For instance the term serofibrinous pleurisy implies that the effusion is serous empyema implies the presence of pus in the pleural cavity pneumothorax the presence of air and hemothorax the presence of blood

The wet and the dry types of pleurisy do not have etiologic implications for in reality they may be but phases of the same disease These terms wet and dry are em ployed to designate the condition of the pleura at a particular time As pleurisy runs its course at one time or another it may pass through both the wet and the dry phases Fibrinous or dry pleurisy is the beginning phase of most pleurisies It is the phase before an effusion into the pleural cavity takes place After effusion dry pleurisy ceases to exist and there is present pleurisy with effusion If the effusion contains bacteria and is thus infected the condition is empyema

Dry Pleurisy During the course of many diseases pleurisy without a significant amount of fluid (less than 500 ml) in the pleural cavity may occur This is the so called dry pleurisy Dry pleurisy often is associated with the beginnings of pneu monia less frequently with pulmonary abscess, gangrene carcinoma or infarctions or with septicopyemia rheumatic fever or chronic nephritis or it originates by extension *from pericarditis peritonitis or hepatitis It has some relation to tubercu* losis occurring sometimes as a primary infection more commonly as a secondary event to a pulmonary tuberculous focus Carcinomatous metastasis to the pleura is soon followed by pleural effusion

The first complaint by the patient who has a pleurisy is stitch pains in the side usually in the neighborhood of the nipple increased by movement and especially by inspiration and accompanied by a dry and painful cough Both cough and respira tion are restrained and the patient bends toward the affected side in order to mini mize the pain For the *same reason the breathing is hurried shallow jerking* and mainly abdominal in type Fever is usually present but the temperature seldom exceeds 101 F (38 3 C) and in mild illness may hardly rise above the normal There *may be a moderate leukocytosis*

On examination of a patient who has pleurisy there may or may not be a palpable friction fremitus On auscultation a friction rub of variable intensity can be heard The sound is audible during both expiration and inspiration The pain and sounds are both increased on pressure over the affected part by the stethoscope or examining hand Hiccough is common Care should be exercised in order to differentiate the sounds of a dry pleurisy from those caused by the skin slipping under the stethoscope a muscle sound an atelectatic crackle and hair on the chest A dry pleurisy *may* disappear in a few days or it *may* last for weeks or even for months

The diagnosis is established from the foregoing findings and from roentgeno *logic examination*

Wet Pleurisy (Pleurisy With Effusion) Pleurisy with effusion usually follows acute pleurisy There is inflammation of the pleura associated with a fluid exudate The fluid may be serous serofibrinous serohemorrhagic purulent or *putrid*

SYMPTOMS The general symptoms of pleurisy may not be pronounced The fever is rarely high it may be continuous or remittent and the temperature usually falls by lysis In some younger patients and often in the aged the onset of pleurisy is insidious There may be no complaint at all or the patient may complain of nothing more than a little shortness of breath and on physical examination the true nature of the illness is discovered The most constant general symptom of pleurisy is pain It is sometimes severe at the beginning and is increased by any movement especially by deep inspiration coughing or sneezing As soon as enough effusion develops to separate the surfaces of the pleura the pain ceases The pain may be referred to the back or the thorax to the shoulders and arms and occasionally into *the abdomen The pain may erroneously be thought due to* intercostal neuralgia muscular rheumatism or an acute surgical condition within the abdomen Chills or chilly sensations are common and sweats occur especially during remissions of the fever and at *night Digestive disturbances including anorexia* and nausea and vomit ing are more constantly present in those who have chills and fever than in those who are afebrile If the effusion develops rapidly there may be a decrease in the amount of urine However *it is more likely that the decreased amount of urine is* the result of the vomiting and inability to take a sufficient amount of fluid by mouth When the pleural exudate is being absorbed it rarely can be absorbed fast enough to increase the output of the urine

EXAMINATION When the patient is examined it is noticed that the posture may be characteristic The patient has less pain usually if he lies on the affected side and assumes a diagonal position so as to give more freedom for *expansion* of the

lung on the healthy side If there is an extensive effusion there may be orthopnea and the sitting position is therefore assumed When the effusion has developed fully the scapula on the diseased side stands a little high and there is a slight lateral curvature of the spinal column with concavity toward the healthy side The skin over the affected region may be stretched On inspiration there is a lagging on the diseased side and the expansion is less The intercostal spaces may be widened and Litten s phenomenon is absent The veins of the neck are distended and the apex beat of the heart is visible and can be seen displaced toward the side opposite the effusion On palpation there are often changes in vocal fremitus However an interpretation of vocal fremitus in pleurisy with effusion is difficult If the effusion reaches any considerable size on percussion there is dullness or flatness over the effusion The upper limit of flatness and increased resistance on percussion assumes a typical curve the upper part of the curve reaching its highest level at the posterior axillary line whence it curves downward and forward toward the middle line in front This line sometimes takes the form of a letter S turned on its side The effusion may fill a large part of one pleural cavity then there is dullness over the whole side of the thorax from base almost to apex One of the most important signs to be found in examining a patient who has pleurisy with effusion is the increasing intensity of the dullness or flatness or a feeling of resistance on percussing from above downward with the finger tips

On auscultation of the thorax occasionally there are distant breath sounds or bronchial breathing bronchophony and often rales may be heard Accentuation of the breath sounds on the normal side is theoretically present but practically this distinction usually cannot be made Egophony is rarely heard

Diagnosis The diagnosis of pleurisy with effusion is confirmed and often made by a roentgenographic examination Small effusions (less than 500 ml) which can not be determined on physical examination always are revealed in roentgenograms of the thorax If roentgen rays are not available it is advisable to make an exploratory puncture when pleurisy with effusion is suspected to exist There are several atypical forms of pleurisy with effusion which may exist as circumscribed pockets along the diaphragm or the interlobar portions of the lung or the disease may be largely confined to the pleura along the mediastinum In any of these atypical forms of pleurisy with effusion when there are no symptoms of pleurisy exploratory puncture should always be considered whether or not the pleurisy is a part of a so called polyserositis In obscure infections the possibility of a small empyema which can not be differentiated from pleurisy with an effusion in any other way except by the exploratory needle makes this diagnostic procedure imperative

Bacterial Infections of the Pleura ***Empyema*** There are many bacteria which are capable of producing empyema Pleural infection is very commonly associated with lobar pneumonia and other pneumococcal infections These organisms gain access to the pleural cavity by extension from the lungs Likewise tuberculous empyemas occur in pure form or as mixed infections

Empyema may occur during the course of septicemia The *Escherichia coli* and *Salmonella typhosa* and paratyphi groups of organisms may invade the pleural cavity particularly if there is peritonitis from these organisms *Pasteurella tularensis* and *Brucella* have been found in empyemic fluid

In still other instances streptococcal staphylococcal and pneumococcal empyemas and septicemia may follow a pneumonia or a pulmonary abscess caused by these organisms

The hemolytic streptococcus may reach the pleural space by direct extension from streptococcal pneumonia In some instances however the so-called primary empyema is associated with streptococcal bacteremia

Symptoms Pleurisy is often an early manifestation of pneumonia and is evidenced as a stitch and later a pain in the lower part of the thorax The pain is usually

lost during the course of the pneumonia not to recur until after the crisis or lysis and then only if there is a pleural infection as a complication of the pneumonia

Empyema may make its appearance during the course of pneumonia Often the febrile course of the pneumonia is unduly prolonged The patient continues to show evidence of toxemia as manifested by weakness and marked leukocytosis all of which are not evident until the course of the illness is obviously too prolonged for pneumonia

In the usual instance however the empyema occurs after the pneumonia has run its course The patient s temperature and leukocyte count have reached normal limits After one to two weeks of seemingly satisfactory convalescence fever again appears accompanied by profuse sweating There may or may not be pleural pain The leukocyte count increases and a mild secondary anemia is present

In those who have both pneumonia and bacteria which precede the pleural infection the diagnosis of pyemia may have already been made These patients are critically ill and the mere presence of empyema is of no importance so far as the outcome is concerned

When a large quantity of foul smelling pus is expectorated there is an extension of an empyema to form a bronchial fistula (putrid empyema)

Examination On examination of the thorax by physical means there may be a sufficient amount (500 ml) of pleural effusion present to produce characteristic physical findings More frequently however there is too small an amount of pus present to be definitely detected by physical examination

In other instances the signs of pleural effusion are masked by an underlying pulmonary consolidation

In instances in which surgical treatment is contemplated the introduction of 10 ml of heavy iodized oil such as Lipiodol is a useful maneuver for determining the size and the most dependent portion of the empyematic cavity The heavy oil sinks to the most dependent portion of the empyema pocket Postero anterior and lateral films made with heavy exposure and with the patient in the upright position will indicate the optimal point at which to secure dependent drainage of the cavity

If there have been evidences of rupture of an empyemic cavity into bronchus such as the expectoration of quantities of pus a bronchopleural fistula may be present The bronchopleural fistula may be demonstrated by the injection of 2 ml of a 1 per cent aqueous solution of gentian violet at the time of the diagnostic thoracentesis If a fistula exists the dye will appear in the sputum within 24 to 48 hours after injection

Diagnosis The diagnosis is usually made on the basis of roentgenologic examination A pleural exudate following pneumonia requires thoracentesis to distinguish between empyema and the sterile effusion which occurs after lobar pneumonias The aspirated fluid is cultured bacteriologically to demonstrate with certainty the nature of the causative organism The effusion in instances of lobar pneumonia is usually sterile When the effusion is sterile no treatment is necessary

Encapsulated empyema occurs along the pleural surfaces usually between the lobes of the lungs The presence of an encapsulated empyema can be established only by roentgenologic examination and aspiration of the encapsulated pus

Complications of Empyema Infection of the thoracic wall is more likely to develop in those who have large abscess of the lung with profuse external discharge in the putrid type of empyema

Empyema necessitatis is the spontaneous rupture of an empyema through the parietal pleura into the thoracic wall Eventually the pus breaks through the skin This condition is rarely encountered except with tuberculous and actinomycotic infections

A bronchopleural fistula may make its way to the surface of the thoracic wall

or it may drain into the bronchial system Occasionally an empyema may drain completely by this route and heal spontaneously

Septicemias and pyemias may either precede or follow empyema

Tuberculosis of the Pleura The tuberculous pleurisies occur in (1) those who have recognizable pulmonary tuberculosis and (2) those in whom pulmonary tuberculosis is not recognized There may or may not be a pleural effusion

Two factors according to Meyers seem to be necessary for the development of a tuberculous pleural effusion (1) An individual must be sensitive to tuberculin by having or having had tuberculosis (2) The tuberculin must reach the pleural space The tuberculin probably reaches the pleural space by the rupture of a small caseous pleural tubercle

When a subpleural tuberculous focus enlarges and involves the pleural membrane pleuritis ensues A firm scar binds the visceral and parietal pleurae forming a pleural adhesion Occasionally with the onset of pleural inflammation there is a rapid accumulation of serofibrinous exudate in the pleural space and if the quantity of this exudate is sufficient (500 ml or more) a detectable clinical pleural effusion is present

SYMPTOMS The general symptoms of pleurisy have been described in discussions of the various forms of pleurisy Dry pleurisy is manifested by pain on respiration The pain is well localized and is aggravated by deep breathing coughing and movements of the thorax The pain from involvement of the diaphragmatic pleural surface is often referred to the posterior midshoulder region of the affected side The pain often begins abruptly and increases rapidly in intensity

There may or may not be fever When present fever is irregular in its appearance and is rarely high It increases as effusion takes place and in extremely acute tuberculous pleurisy the temperature may reach 105 F (40 5 C) If a large effusion accumulates symptoms of increasing intrathoracic pressure appear

EXAMINATION Physical examination may reveal reduction of respiratory excursion in the involved half of the thorax A pleural friction rub or fine dry pleural crackles are frequently heard

On development of pleural effusion the friction rub disappears to be replaced by the signs of fluid dullness to flatness on percussion reduction or absence of breath sounds voice sounds and tactile fremitus and egophony The trachea and other mediastinal structures may be shifted away from the side of the effusion the degree of displacement depending on the amount of fluid present and the mobility of the mediastinum

In tuberculous pleurisy except during the very acute manifestations of the disease the total number of leukocytes is usually within the normal range During the very acute phase of the disease there is a polymorphonuclear leukocytosis which quickly subsides

A tuberculous effusion when withdrawn is a straw colored fluid Leukocytes in the fluid ordinarily number 500 to 3 000 per cubic millimeter nine tenths of which are lymphocytes The fluid may contain blood

Guinea pig inoculation or culture on special media for tubercle bacilli should be a routine procedure but by all methods of examination the results in 30 to 50 per cent of these fluids are negative for tubercle bacilli

DIAGNOSIS Roentgenologic examination demonstrates evidence of the pleurisy Tuberculous pleuritis may occur in a patient who has evidence of an active lesion in the lung In this instance the diagnosis of tuberculous pleurisy is obvious

Dry tuberculous pleurisy however may and often does exist in the absence of demonstrable pulmonary tuberculosis and without pleural effusion

A negative tuberculin reaction which remains negative for a few months after the attack is good evidence against a tuberculous etiology

If a large effusion is present when the patient is first seen a roentgenologic

lost during the course of the pneumonia not to recur until after the crisis or lysis and then only if there is a pleural infection as a complication of the pneumonia

Empyema may make its appearance during the course of pneumonia Often the febrile course of the pneumonia is unduly prolonged The patient continues to show evidence of toxemia as manifested by weakness and marked leukocytosis all of which are not evident until the course of the illness is obviously too prolonged for pneumonia

In the usual instance however the empyema occurs after the pneumonia has run its course The patient s temperature and leukocyte count have reached normal limits After one to two weeks of seemingly satisfactory convalescence fever again appears accompanied by profuse sweating There may or may not be pleural pain The leukocyte count increases and a mild secondary anemia is present

In those who have both pneumonia and bacteria which precede the pleural infection the diagnosis of pyemia may have already been made These patients are critically ill and the mere presence of empyema is of no importance so far as the outcome is concerned

When a large quantity of foul smelling pus is expectorated there is an extension of an empyema to form a bronchial fistula (putrid empyema)

EXAMINATION *On examination of the thorax by physical means there may be a* sufficient amount (500 ml) of pleural effusion present to produce characteristic physical findings More frequently however there is too small an amount of pus present to be definitely detected by physical examination

In other instances the signs of pleural effusion are masked by an underlying pulmonary consolidation

In instances in which surgical treatment is contemplated the introduction of 10 ml of heavy iodized oil such as Lipiodol is a useful maneuver for determining the size and the most dependent portion of the empyematic cavity The heavy oil sinks to the most dependent portion of the empyema pocket Postero anterior and lateral films made with heavy exposure and with the patient in the upright position will indicate the optimal point at which to secure dependent drainage of the cavity

If there have been evidences of rupture of an empyemic cavity into bronchus such as the expectoration of quantities of pus a bronchopleural *fistula may be* present The bronchopleural fistula may be demonstrated by the injection of 2 ml of a 1 per cent aqueous solution of gentian violet at the time of the diagnostic thoracentesis If a fistula exists the dye will appear in the sputum within 24 to 48 hours after injection

DIAGNOSIS The diagnosis is usually made on the basis of roentgenologic examination A pleural exudate following pneumonia requires thoracentesis to distinguish between empyema and the sterile effusion which occurs after lobar pneumonias The aspirated fluid is cultured bacteriologically to demonstrate with certainty the nature of the causative organism The effusion in instances of lobar pneumonia is usually sterile When the effusion is sterile no treatment is necessary

Encapsulated empyema occurs along the pleural surfaces usually between the lobes of the lungs The presence of an encapsulated empyema can be established only by roentgenologic *examination and aspiration of the encapsulated pus*

COMPLICATIONS OF EMPYEMA Infection of the thoracic wall is more likely to develop in those who have large abscess of the lung with profuse external discharge in the putrid type of empyema

Empyema necessitatis is the spontaneous rupture of an empyema through the parietal pleura into the thoracic wall Eventually the pus breaks through the skin This condition is rarely encountered except with tuberculous and actinomycotic infections

A bronchopleural fistula may make its way to the surface of the thoracic wall

the subsequent tuberculous empyema may be limited but a mixed infection may develop later Most of the patients in whom spontaneous pneumothorax develops in the course of tuberculosis become seriously ill All of them have fever and without exception a pleural exudate appears soon after the onset About half the patients die within one month and probably fewer than 1 in every 10 regain health

EXAMINATION On examination of a patient who has a pneumothorax the affected side may be distended and nearly or absolutely immobile on respiration The apex beat of the heart is displaced toward the normal side unless the mediastinum is immobile The vocal fremitus is enfeebled or absent The trachea may be displaced toward the normal side The percussion note is hyperresonant but it varies according to the tension of the gas in the pleural cavity The affected side may be considered to be normal on percussion while the normal lung is thought to be the one affected On listening at the back of the thorax while someone else taps one coin on another in front a distinct metallic echoing sound is heard If there is a considerable amount of fluid or pus present in the thorax the percussion dullness may be bounded by a horizontal line and this line remains horizontal irrespective of change in posture of the thorax On shaking the patient a metallic splashing sound may be audible When a pulmonary fistula is patent and is below the surface of the fluid in the pleural cavity on the patient s lying down or standing the sound of gurgling fluid may be heard On auscultation over a pneumothorax the breath sounds may vary from markedly feeble to loud and amphoric

DIAGNOSIS If no abnormalities are found on physical examination the roentgenologic examination will reveal the presence of pneumothorax Fluid accumulates rapidly in the pneumothorax pocket It is important to determine whether the bronchopleural communication remains open for if there is a widely open bronchopleural communication withdrawal of large volumes of air does not alter the intrapleural pressure

The early management of these patients is concerned with removal of the pneumothorax air as needed In the mildest forms of the disease treatment may not be necessary

Rheumatic Pleurisy Rheumatic fever causes a small number of cases of serofibrinous pleurisy with effusion

Rheumatic fever attacks serous membranes selectively for instance the synovial membranes the endocardium and the pericardium are common sites of rheumatic infection In a few cases the pleural membrane also is affected with the production of a fibrinous pleurisy often quickly followed by serous pleural effusion This sequence of events is particularly likely to occur in the presence of rheumatic pneumonitis or pericarditis whence the infection apparently spreads directly or by extension along the lymphatics

Those who have rheumatic pleurisy have had rheumatic fever and in all but a very few the pleural involvement occurs in the course of an acute exacerbation of symptoms The pleuritis may be preceded in onset by polyarthritis However in some patients the joints become involved after the onset of the pleurisy The symptoms in this latter group of patients are those of rheumatic polyarthritis accompanied by the development of pleuritic pain Characteristically the patient is very ill with high fever and dyspnea Rheumatic pleurisy often co exists with pericarditis Rarely a subacute or chronic rheumatic pleuritis is observed

EXAMINATION Early in the course of rheumatic pleurisy a pleural friction rub may be heard but this is quickly superseded by signs of effusion In almost all of these patients examination by physical means reveals evidence of rheumatic mitral heart disease

A history of previous rheumatic fever or the presence of a rheumatic cardiac lesion is the chief clue in making a correct diagnosis Rheumatic pleurisy is identified when a serous pleural effusion develops in the course of rheumatic fever The

examination is made after thoracentesis A definite diagnosis of tuberculous pleurisy is made by demonstration of the tubercle bacillus by inoculation of a guinea pig

Histologic studies of the cells in the sediment should be made for presence of *malignant cells*

Tuberculous Empyema Tuberculous empyema exists if the pleural exudate is definitely purulent and contains tubercle bacilli in sufficient numbers to be demonstrable *in an unconcentrated smear A tuberculous empyema is termed simple* when the tubercle bacillus is the sole infecting organism, and *mixed* when in addition one or more other types of pathogenic bacteria are present in the pleural cavity

1 ***Simple Tuberculous Empyema*** The visceral and parietal pleura are considerably thickened by fibrous tissue together with caseous material The pleural pus contains only tubercle bacilli which are found by direct smear examination

Tuberculous pleural empyema may occur spontaneously but often is present in those who have received artificial pneumothorax for pulmonary tuberculosis

The pathologic process is a subpleural tuberculous focus which has spread by caseation liquefaction and rupture into the pleural cavity

At the onset of the empyema there is some fever and often pain in the chest The acute empyema may come on abruptly usually soon after the induction of pneumothorax owing to extensive pleural caseation *In these instances the temperature* rises rapidly to 104 F (40 C) and chills occur Pleural pain in the involved side is an early symptom The fever may persist for several weeks and subside *gradually Between these mild and severe attacks of the disease gradations* in severity of the clinical manifestations may exist

The mortality rate in all cases of simple tuberculous empyema has been high from 30 to 40 per cent

The diagnosis of simple tuberculous empyema is made when the presence of fluid in the pleural cavity is determined and aspiration yields a turbid or purulent exudate containing acid fast bacilli on direct culture or smear but giving sterile cultures for other bacteria

2 ***Mixed Tuberculous Empyema*** In the mixed tuberculous empyema the exudate is purulent and may contain a wide mixture of organisms

The pleural infection arises from a rupture of a caseous focus *in the visceral* pleura of a tuberculous lung with the production of a bronchopleural fistula When such an event occurs tubercle bacilli as well as the pyogenic flora of the bronchial *tree reach the pleural cavity in large numbers*

The symptoms of the mixed tuberculous empyema are the same as those of simple tuberculous empyema but more severe

The presence of a mixed infection may not be suspected until cultures of the pus reveal the pyogenic organisms The diagnosis of tuberculosis is suspected and proper isolation and care of the patient have been executed long before the presence of tubercle bacilli has been demonstrated by cultures

The prognosis is grave more than 70 *per cent mortality is to be expected*

Tuberculous Spontaneous Pneumothorax A region of tuberculous caseation originates in the subpleural tissues and extends to the pleural membrane liquefies *it and ruptures* into the pleural space When rupture takes place air tubercle bacilli and other microorganisms from the bronchial passages are admitted to the pleural cavity

SYMPTOMS The patient who is known to be suffering from pulmonary tuberculosis has a sudden pain in the chest becomes dyspneic and cyanotic and experiences varying degrees of shock In those who survive the initial episode chills and fever ensue

In a few cases the pleural *opening is small* and closes rapidly When this occurs

which leave a moderate pleural fibrosis which persists At times pleural effusions alone occur which are absorbed leaving no evidence of their presence

The findings on examination depend on the presence or absence of pneumonitis and pleural effusions

Diagnosis depends on the status of the lung and pleura before roentgen therapy If the examinations of the lung and pleura were recorded before roentgen therapy and during the therapy if no other reason for the pleural changes occurred it may be assumed that the pleurisy resulted from the irradiation therapy

Traumatic Pneumothorax Direct violence may cause pneumothorax by (1) puncture of the lung by the sharp end of a fractured rib (2) blows and crushing injuries without puncture of the thorax in these injuries there are rupture of the visceral pleura and intrapulmonary alveolar tears The air then makes its way along the bronchovascular bundles to the hilus and reaches the mediastinum causing mediastinal emphysema Secondary rupture of the mediastinal parietal pleura then produces pneumothorax (3) penetrating wounds and (4) artificial overinflation of the lung The use of excessive positive pressure in intratracheal anesthesia and pulmotors can produce pneumothorax

In most of the instances caused by trauma the amount of air entering the pleural space is moderate or small However massive tension pneumothorax is occasionally seen

Symptoms Often there is shock and this masks all other symptoms If there is no shock pain in the injured side of the thorax is severe Dyspnea and cyanosis are severe if the pneumothorax is large In those who have small partial collapse no symptoms in addition to those of the injury are observed

Examination Physical examination in pneumothorax of moderate or large size reveals hyperresonance to percussion which is often not great The breath sounds are reduced or absent over the involved side of the thorax When a pneumothorax is extensive the heart and trachea are frequently found to be displaced away from the side of the collapse Signs of fluid in the pleural cavity are due to intrapleural blood from the site of injury Subcutaneous emphysema is a common occurrence in traumatic pneumothorax It usually develops when a break in continuity of the parietal pleura allows the escape of air from the pleural space which then makes its way to the subcutaneous tissue and may spread widely over the body If extensive subcutaneous emphysema is painful

Diagnosis The presence of air in the pleural cavity is determined by the combined use of physical and roentgenologic methods The roentgenologic method is the only one by which the degree of collapse can be accurately determined

Chylothorax Chylothorax consists of the accumulation of chyle in the pleural cavity This fluid has the appearance and stability of a solution of homogenized milk It contains varying amounts of fat (2 to 3 per cent) which are determined by the Babcock test as employed by dairymen

Injuries produced by hyperextension of the spinal column may rupture the thoracic duct After the injury a latent period of weeks may pass before fluid appears in the pleural cavity The delay is due to retropleural accumulation of chyle This chyloma appears as a mediastinal mass in the roentgenogram The chyloma ruptures into the pleural space and chylothorax ensues Chylothorax like chyluria may occur spontaneously

Occasionally a mediastinal lymphosarcoma or Hodgkin s disease may completely obstruct and rupture the thoracic duct and produce chylothorax

The development of some instances of chylothorax is characteristic The onset of pleural pressure symptoms such as dyspnea cyanosis tachycardia and a sense of tightness in the chest is severe Associated with these symptoms is a decline in weight due to the loss of large amounts of digested and absorbed fat In some instances the wasting and exhaustion of the patient continue to progress and the

fluid removed by thoracentesis is an exudate which clots rapidly It is sanguineous in appearance in more than half the cases Leukocytes in the fluid number from 500 to 5 000 per cubic *millimeter and a differential count usually shows* a very slight preponderance of polymorphonuclear cells over lymphocytes Cultures of the exudate are sterile This condition must be differentiated from other pleurisies by the presence of other symptoms or signs of rheumatic disease

PLEURISY DUE TO FILTRABLE VIRUSES

Epidemic Pleurodynia (Epidemic Myalgia) This disease is characterized by an acute febrile illness of brief duration whose chief symptom is severe thoracic pain indistinguishable from that of pleurisy The disease has occurred in small sporadic epidemics which usually are observed during the warm months of the year The incidence is chiefly among children and young adults

From stored throat washings from those who had pleurodynia Miller and associates of Harvard University and Tufts College recently isolated a virus antigenically related to the Connecticut 5 virus previously isolated by Melnick from a case of nonparalytic poliomyelitis

Epidemic pleurodynia has an abrupt onset with fever accompanied by a chill Pain on breathing or coughing appears simultaneously and is limited to the lower intercostal regions of one side However instances with bilateral pain headache abdominal pains and stiffness of the back and neck have been recorded

The temperature reaches 101 to 104 F (38 3 to 40 C) and the fever and pain persist as a rule for 12 to 48 hours Thereafter the symptoms subside rather rapidly but after an interval of 2 days or so about one fifth of the patients have a second paroxysm of pain and fever identical in character with the first but usually somewhat less severe After this recovery is prompt there being no complications Death from epidemic pleurodynia has never been reported

Examination often reveals that the patient has a moderately red throat but *cough is not common The involved side of the thorax may reveal muscle guarding* and there is tenderness on pressure in the intercostal spaces over the painful region Pleural friction rubs on auscultation are inconstantly present

Diagnosis The course of the disease and its occurrence as part of an epidemic must decide the diagnosis It would be impossible to identify a sporadic case of epidemic pleurodynia Leukocyte counts reveal a mild leukopenia during the active stage of the disease

DISEASES OF THE PLEURA DUE TO TRAUMA OR PHYSICAL AGENT

Traumatic Serofibrinous Pleurisy Serofibrinous pleurisy may develop as a sequel of direct trauma to the thorax In general it appears that the serofibrinous pleurisy most commonly is associated with a subpleural hematoma The common causes of serofibrinous pleurisy are fractures of ribs with the formation of hematoma subpleurally at the site of the fracture with or without actual penetration of the parietal pleura by the fractured fragments of bone However hematomas may occur from contusion of the thoracic wall without rib fracture Likewise contusion of a lung due to either direct or contrecoup violence with hematoma beneath the visceral pleura may be associated with serofibrinous pleurisy It seems that the size of the effusion is ordinarily small and the fluid is blood stained Cultures of these effusions are sterile unless secondarily infected by pyogenic bacteria

Pleurisy Following Irradiation Therapy After intensive roentgen therapy to the thorax a pulmonary reaction may occur In the beginning there is the appearance of a pneumonitis which progresses slowly resolves slowly and heals poorly for sometimes there is left an increase in fibrous tissue in the involved region of the lung In some cases of this roentgen pneumonitis changes develop in the pleura

Pleural Transudates *Venous Passive Congestion* The usual cause of passive congestion in the veins draining the visceral and parietal pleura is congestive cardiac failure Constrictive pericarditis has a somewhat similar effect In the occasional case of obstruction of the superior vena cava congestion of the parietal pleura develops because of interference with return flow through the azygos system

The symptoms are those of congestive heart failure The findings on examination by physical means are those of heart failure and fluid in the pleural space especially on the right side of the thorax

The fluid obtained by thoracentesis is a thin straw colored fluid relatively clear and having a specific gravity usually less than 1 014 If the effusion has been present for some time reabsorption of water may cause concentration of the proteins giving a higher specific gravity In uncomplicated cases the fluid contains very few cells usually less than 500 per cubic millimeter and these are lymphocytes The presence of polymorphonuclear leukocytes and erythrocytes suggests the existence of a complicating pulmonary infarct or a secondary infection of the transudate an empyema An empyema in a patient who has heart failure is suggestive of a dispersion of infected thrombi from a bacterial endocarditis

PLEURISY OF ENDOCRINE ORIGIN

(Meigs Syndrome)

In the presence of benign fibroma of the ovary there may be associated a sterile serous pleural effusion and serous ascites Women between the ages of 30 and 65 years complain of dyspnea and unilateral thoracic pain There is no fever Examination reveals physical signs of pleural effusion the presence of which is confirmed by the roentgenogram The ovarian fibroma is variable in size but can ordinarily be detected on pelvic examination

Thoracentesis yields a thin straw-colored fluid which contains but few leukocytes—usually fewer than 1 000 per cubic millimeter—the majority of which are lymphocytes Cultures and inoculation of guinea pigs with the fluid are negative Results of examination for neoplastic cells are negative

The diagnosis is confirmed by the finding of fibroma of the ovary in the absence of heart failure or embolism A patient who is found to have an ovarian tumor with pleural effusion everything else being negative for positive evidence of malignant disease should have the pelvis surgically explored if there be an enlargement of an ovary Patients afflicted with Meigs syndrome have recovered completely after removal of the ovarian fibroma

SPONTANEOUS PNEUMOTHORAX

The term spontaneous pneumothorax implies that air has spontaneously entered the pleural space and thus produced a collapse of the lung Spontaneous pneumothorax despite its cognomen in the series of pneumothoraces usually results from rupture of an emphysematous bulla situated on the surface of the lung into the pleural space

The normally existing negative intrapleural pressure is an expression of the physiologic internal elasticity of the lung which brings it to a smaller volume while at rest The lung remains expanded because of its internal atmospheric pressure which is a greater force than this normal force of contractility of the lung When a communication develops between the pleural space and the atmosphere through an opening in the thoracic wall the atmospheric pressure in the alveoli is permitted to equalize itself with the atmospheric pressure The normal pulmonary elasticity contracts the lung to a fraction of its normally expanded volume Air is therefore permitted to enter the pleural cavity to compensate for the shrinkage and pneumothorax results In an otherwise normal thorax the difference in pressure between the two pleural cavities causes a displacement of the mediastinal structures away from the side of the pneumothorax

condition is fatal within a few weeks In other cases *after the initial accumulation* of pleural fluid the mediastinal pressure becomes stabilized so that pressure symptoms are no longer severe The pressure appears to reduce the leakage of the fluid into the pleural sac

Some patients carry a large chylothorax and enjoy fairly good health However most of those who have chylothorax die within a period of a few months

Physical and roentgenologic signs are those of pleural effusion The presence of chylothorax is established by thoracentesis Aspiration yields a fluid having the appearance of milk The Babcock test for fat reveals a fat content of from 1 to 2 *per cent or more which establishes the diagnosis*

Hemothorax In hemothorax the pleural cavity contains blood little if at all diluted with serous exudate Hemothorax originates from many causes trauma and malignant disease are the common agents

After an intrapleural hemorrhage has taken place the blood gradually undergoes a change in color (conversion of hemoglobin to bilirubin) If the blood withdrawn by thoracentesis is centrifuged a yellow or brownish tinge in the supernatant plasma indicates that bilirubin has already been formed and therefore the hemorrhage must have begun at least a day or two or longer prior to the aspiration

Hemopneumothorax Hemopneumothorax is the combined presence of blood and air in the pleural cavity This condition is most frequently the sequela of traumatic pneumothorax and the blood comes from an intrapleural wound

Hemopneumothorax however occasionally occurs in idiopathic pneumothorax and more rarely in the tuberculous and emphysematous types of spontaneous pneumothorax If the amount of blood in the thorax is small it will not affect the course of the pneumothorax

A low grade fever may be produced by pleural irritation and disintegration of blood In the rare instances in which severe hemorrhage has occurred shock may ensue

Except in those known to have tuberculosis hemopneumothorax is suspected when a sizable effusion appears in the pleural cavity of a person who has a pneumothorax Aspiration of some of the fluid confirms the diagnosis

The blood is removed from the pleural space to prevent future fibrosis and calcification

PLEURAL EFFUSIONS SECONDARY TO DISORDERS OF CIRCULATION

These effusions are usually sterile like those from the pneumococcus and rheumatic pleurisies Pneumococcal empyema occurs in those who have had *untreated lobar pneumonia*

Pleural Effusion Due to Pulmonary Infarction The occurrence of a pulmonary infarct is followed by an intense inflammation like reaction of the pleura *when the infarct is situated near the periphery of the lung* A thin layer of fibrinous exudate rapidly appears on the visceral pleura overlying the infarct Then a serous exudate begins to accumulate in the pleural space the fluid developing *rapidly and commonly becoming sanguineous* from leakage of erythrocytes from the intensely congested infarcted tissue

Immediately after the infarct there may be no signs of pleurisy on examination by *physical means* Twelve to 24 hours after the accident a pleural rub and later the signs of pleural effusion will be present in all subpleural infarctions

The diagnosis is established by roentgenoscopic examination In case the pleural *exudate is examined* it will reveal a leukocyte count in the fluid of 1 000 cells per cubic millimeter or less The fluid is sterile on culture and contains no tumor cells and tubercle bacilli cannot be recovered by guinea pig inoculation

PLEURAL THICKENING

During the routine examinations of the thorax there may not be signs of pleural thickening Roentgenologic examination may however reveal some thickening of the pleura These pleural thickenings are of varying degrees and extent A chronic inflammation of the pleura with formation of new fibrous tissue and with adhesions between two layers in rare instances may extend to the point of obliteration of the whole pleural cavity on one side with retraction of the thorax The side of the thorax which is affected is shorter than the other There is scoliosis with concavity toward the shrunken side The shoulder on the affected side is depressed and the intercostal spaces are narrowed There are diminished expansion dullness on percussion and enfeeblement of the breath and voice sounds and of vocal fremitus Litten s phenomenon is absent As the side becomes shrunken the mediastinum and the heart are drawn toward it If there be much thickening an exploratory needle may have to be jabbed through a thick layer of tough fibrous tissue In some instances of this type of pleurisy the whole pleura becomes calcified Adhesions to the diaphragm are often exquisitely demonstrated on roentgenographic examination

Commonly however there will be observed circumscribed adhesions only usually to the diaphragm or at the bases of the lungs The etiology of these pleural thickenings is not known Great thickenings of the pleura are most often due to tuberculous pleurisy

If signs and symptoms of this type of pleurisy are present they usually are minimal At times however the patients may be slightly cyanotic and they often have some dyspnea on exertion

MALIGNANT TUMORS OF THE PLEURA

Primary Malignant Lesions The pathologists have complained of the difficulty in the histologic classification of primary pleural malignant tumors These tumors may be called carcinomas sarcomas endotheliomas and mesotheliomas The manifestations of these tumors are not distinctive nor can the neoplasms be definitely separated from secondary metastatic malignant lesions

The metastatic tumors which involve the pleura are mostly carcinomas and sarcomas

The first symptom of pleural tumors is heaviness or fullness in the thorax often a dull aching sensation Soon pain is associated with respiration and it is sharp and cutting in nature Dyspnea shortness of breath cough expectoration and loss of weight and strength follow in rapid sequence

Fever palpitation and tachycardia are common The evidence of cardiac embarrassment through mediastinal compression which may ensue is the presence of edèma of the extremities and ascites Ptosis enophthalmus and contracted pupil (Horner s syndrome) may result from pressure on autonomic nerves

In some instances on examination there is a tumor of the thoracic wall Decreased expansion of the thorax edema of the arms and legs abdominal ascites engorgement of vessels of the thoracic wall and edema may have developed as a result of mediastinal compression and encroachment on the heart or large blood vessels

On percussion of the thorax either dullness or flatness will be elicited over irregular regions Often the neoplasm is diffuse or if there is fluid in the pleural cavity dullness and flatness may be extensive A high diaphragm on the side of the lesion usually can be elicited by physical means and indicates direct diaphragmatic or phrenic nerve involvement Auscultatory findings consist of diminished or absent breath sounds Rarely will there be rales and pleural friction rubs are unlikely

Occasionally as a lung collapses the consequent contraction of the pleural membrane will close the bronchopleural opening and thus halt the process at this point The normal pressure relationships are somewhat re established and the intrapleural pressure remains slightly negative as compared to the atmosphere

The term tension pneumothorax indicates the building up of an intrapleural pressure greater than atmospheric pressure This serious condition is permitted to develop by a bronchopleural communication a one way valve action which allows air to pass only from the lung to the pleural space (Meyers)

Pulmonary Emphysematous Spontaneous Pneumothorax In pulmonary emphysema there are often subpleural bullae containing air and communicating *with the bronchial tree The visceral pleura is the outer wall of the blebs* Rupture of one of these subpleural bullae produces spontaneous pneumothorax In these instances collapse of the lung is only partial In cases of tension pneumothorax the increased intrapleural pressure is great enough to collapse even an emphysematous lung

The patient who has emphysema suffers from severe pleuritic pain in one side Dyspnea and cyanosis are evident except in the mildest instances of pneumothorax and vary in degree with the extent of the collapse Fever is uncommon and pleural effusion usually does not occur

Physical examination reveals the signs of pneumothorax on the side of the pain When subcutaneous emphysema appears it is ordinarily secondary to mediastinal emphysema hence the presence of air is first observed in the tissues of the neck from which it may spread The diagnosis is established by the finding on physical and roentgenologic examination of pneumothorax in a patient suffering from pulmonary emphysema

Spontaneous pneumothorax due to emphysema often recurs There being no pleural inflammation at the time of the collapse adhesions frequently do not form and the same or a different emphysematous bleb may rupture again In some instances multiple recurrences have been observed In a patient who has extensive emphysema severe spontaneous pneumothorax may be fatal

Idiopathic Spontaneous Pneumothorax The term idiopathic pneumothorax is aptly applied because in the rare fatal instances of the disease necropsy has not disclosed a bleb or the point of pleural rupture If a pleural rupture is demonstrated it is the so called subpleural vesicle a local congenital weakness of the pleural tissues

Idiopathic pneumothorax occurs in young healthy adults The onset is with a sudden violent pain in one side of the thorax There may be shock *The respirations* are shallow and rapid The patient may have a slight fever for 2 or 3 days but none thereafter and in many cases the temperature remains normal throughout

Physical examination reveals signs of pneumothorax on the side of the pain and in marked instances some degree of mediastinal displacement away from the side of the lesion

In idiopathic benign spontaneous pneumothorax pleural fluid usually does not appear In some instances a small amount of serous exudate develops If a large pleural effusion is found the most likely cause is hemothorax as a result of bleeding from the lung at the time of pleural rupture

Subcutaneous emphysema occurs occasionally in idiopathic spontaneous pneumothorax The subcutaneous air appears first in the neck and subsequently spreads

A diagnosis of idiopathic spontaneous pneumothorax is justified when the condition develops in the absence of trauma in a young patient who has no known preexisting pulmonary disease A roentgenologic examination reveals and confirms the presence of a pneumothorax The extent of a pneumothorax can be determined only by roentgenologic investigation

in bed The pain is situated in the region of the epigastrium or just above the ensiform cartilage The symptoms may be suggestive of gallbladder disease ulcer or *angina pectoris*

The findings on physical examination depend on how large the hernia may be If the hernia is large there is thoracic tympany and borborygmi are heard in parts of the thorax usually occupied by the lungs In small hernias there are no signs on examination

The hernia is revealed by roentgenoscopic examination after the patient has ingested a barium meal In large hernias the diagnosis may be suspected from physical examination or the history or may be found on a routine roentgenogram of the chest

When diaphragmatic hernias are discovered accidentally and are not causing symptoms no treatment is necessary

False Hernia The false diaphragmatic hernias are always due to an arrest of development of the pleuroperitoneal membrane with a congenital absence of a part of the diaphragm Congenital defects of this kind occur much more frequently on the left side than on the right An absence of a part of the left half of the diaphragm permits the abdominal organs such as the stomach and parts of the intestine and occasionally the spleen the pancreas and the left kidney as well as a portion of the liver to enter the perforated pleural cavity In the extremes most of the pleural cavity is occupied by these organs Such hernias vary in extent from this extreme to those with defects in the diaphragm which admit only small parts of the abdominal contents

The symptoms are variable in degree and often none are present

In extreme instances of diaphragmatic hernia tympany on percussion and peristaltic sounds on auscultation on examination of the thorax are diagnostic In those hernias which admit only a small portion of an abdominal viscus into the pleural cavity the diagnosis is established by roentgenoscopic inspection after the patient has ingested a barium meal

INFECTIONS OF THE DIAPHRAGM

Acute Diaphragmatitis Acute diaphragmatitis is usually associated with acute infections situated either in the thorax or in the abdomen such as pneumonia and peritonitis or with general disease of the body as a whole In other instances of acute diaphragmatitis the etiology is usually not known

There is an acute inflammation with swelling of the muscle fibers which may be limited to the muscle or may extend to its serous coverings Fluid may accumulate followed by adhesions and obliteration of the cardiophrenic and costophrenic angles These inflammatory processes may destroy muscle which is replaced by fibrous tissues and thus the diaphragm partially loses its ability to function and appears flat rather than dome shaped on roentgenoscopic examination

The disease may extend however so as to involve the diaphragmatic pleura resulting in pleural effusion and even empyema It may then extend to the visceral pleura to the diaphragmatic peritoneum and to the liver and thus produce symptoms indicating the extension of the disease

The symptoms commence with a chill and fever followed by pain over the costal margins and in the shoulder region of the involved side Often there are pain and soreness across the upper part of the abdomen The pain is intensified on inspiration and change in posture The symptoms are often very much like those of a pneumonia which affects the base of the lung or both lungs

The symptoms disappear within a few days but often recur at irregular intervals Unless the lung is affected there are little or no cough and expectoration There are often a loss of body weight anorexia and constipation

The patient on examination may or may not appear ill Limitation of motion or a partial immobilization of the diaphragm and the lower part of the thoracic

For practical purposes roentgenologic examination often sufficiently limits the diagnostic possibilities The diagnosis as to the type of tumor *is established by* microscopic study Malignant cells may be demonstrated in the bloody pleural fluid obtained on aspiration

DISEASES OF THE DIAPHRAGM

Position of Diaphragm The peripheral projection of the diaphragm adheres closely to the lower borders of the lungs (see pp 536 and 537)

The diaphragm is *elevated* by peritoneal fluid or air or gas in the digestive tract tumors or pregnancy conditions *within the thorax such as atelectasis* exten sive pulmonary fibrosis and adhesions interruption of a phrenic nerve congenital muscular defects and myasthenia gravis

The diaphragm may occupy a *low position* in acute diaphragmatitis phrenic nerve irritation as may occur in encephalitis tetanus tetany and singultus with peritonitis visceroptosis and asthenias pleural fluid pneumothorax emphysema asthma and intrathoracic tumors

The diaphragmatic muscle may be involved by acute infections The pleural surfaces of the diaphragm *often participate in the infections or malignant lesions* of the rest of the pleural surfaces of the thorax The peritoneal surface likewise participates in the malignant disease and infections of the peritoneal cavity

DISEASES OF THE DIAPHRAGM DUE TO PRENATAL INFLUENCES

Congenital Eventration of the Diaphragm This condition is called also elevation dilatation high position insufficiency relaxation of the diaphragm and Petit s disease

Eventration of the diaphragm is often due to a congenital fault which is a failure of the diaphragmatic musculature to develop on one side usually the left side In place of the muscle there exists only a thin membrane The pleural and peritoneal components are intact but in the absence of muscle *the involved part* of the diaphragm yields to pressure

The condition is characterized by an abnormally high position of one half of the diaphragm The high position is permanent and the diaphragm is intact The abdominal viscera are placed high and occupy space which topographically belongs in the thoracic cavity

There may be dyspnea substernal pain discomfort or pain after meals and occasionally nausea and vomiting Usually however there are no symptoms

On examination an increase in the respiratory expansion of the thorax in the region of the costal margins may be observed while the patient stands This increase in inspiratory expansion is made less obvious when the patient is in the recumbent posture Tympany may be elicited over the lower part of the thorax which is nor mally occupied by lung Borborygmi may be heard over the lower part of the thorax in positions where these sounds are not normally audible

The diagnosis is established by roentgenoscopic study after the patient has ingested a barium meal

In most cases of eventration of the diaphragm no treatment is necessary

Congenital Diaphragmatic Hernia Congenital diaphragmatic hernias are separated on an anatomic basis as true hernias and false hernias

True Hernia *In a true diaphragmatic hernia* the abdominal contents extend through the diaphragm into the thorax within the sac formed by the serous cover ings of the diaphragm These hernias occur most frequently at the esophageal foramen However *they may occur in any part of the diaphragm They often result* from congenital imperfections in the diaphragmatic muscle

Often there are no symptoms In most instances of diaphragmatic hernia how ever pain is present when the stomach is *full and often while the* patient is lying

Secondary Diaphragmatitis Pneumonia and peritonitis are common causes of secondary diaphragmatitis In basal lobar pneumonia marked diaphragmatic changes occur and in fatal instances of the disease the postmortem examinations often reveal waxy degeneration of the muscle fibers In cases of bronchial pneumonia the diaphragmatic changes are slight and recovery is often complete

Inflammation of the pleura often extends to the diaphragm A persistent purulent exudate impairs the functions of the diaphragm permanently The diaphragm is most frequently attacked by tuberculosis Tubercles are found within the muscle itself

The weight of pleural effusion damages the diaphragm by producing stretching deformity and loss of elasticity When fluid is present in the pleural space there is a tendency for adhesions to form in the costophrenic angles and between the pericardium and the diaphragm fixing the diaphragm in an abnormal position which impairs its function In cases of extreme diaphragmatic adhesions the heart may be displaced

In any chronic infection of the lung when dyspnea is out of proportion to the amount of pulmonary involvement an impairment of diaphragmatic function is suspected as being at least partially responsible In tuberculosis and pneumoconiosis bilateral diaphragmatic adhesions may result in marked restriction of the diaphrag matic excursion and thus dyspnea is inevitable

SUBDIAPHRAGMATIC ABSCESS

Subdiaphragmatic or subphrenic abscess is a collection of pus beneath the diaphragm It is usually subsequent to some suppurative process within the abdomen Other causes however have included infection carried from some distant focus by the blood or lymph and invasion of the subphrenic space by extension through the diaphragm of a suppurative process within the thorax

The subphrenic portion of the body extends from the diaphragm to the trans verse colon This region is subdivided by the liver and the arrangements of its peritoneal folds or ligaments to form three subphrenic spaces on the right side and one space on the left

LOCATION OF ABSCESS The right side is more frequently involved than the left because the most frequent diseases preceding subphrenic abscess are lesions of the appendix gallbladder and duodenum There are occasions however when these lesions are followed by subphrenic abscess on the left side

The microorganisms found on culture from subdiaphragmatic abscesses are in the order of frequency streptococci gram negative bacilli staphylococci *Escherichia coli* Proteus and various micrococci

SYMPTOMS The symptoms of subphrenic abscess depend on the combination of many variable factors and are therefore largely dependent on the symptoms of the original lesion or on the postoperative discomforts incident to a recent operative procedure These discomforts may accentuate diminish or completely mask the symptoms produced by the subphrenic abscess The particular sub phrenic space involved by the abscess the onset of the disease the general condition of the patient the presence or absence of complications and the nature of those complications present will influence the symptoms The symptoms may be sum marized as general reactions local reactions and pleural reactions

The general reaction often is that of overwhelming infection The onset is sudden with fever chill and fever and the patient is seriously ill When the infection becomes localized there is pain situated in the region of the diaphragm and the findings may simulate a pleurisy

The pleural and local manifestations revealed by physical examination are often meager indeed despite the presence of intense pain with high fever However in some there is evidence of pleural effusion or pneumonia

wall may be observed In the beginning there are no abnormal auscultatory findings *There is no descent of the diaphragm on inspiration* A pleural effusion may be present

As the disease develops and the diaphragm becomes immobile there may be congestion at the bases of the lungs Spasm of the transversus abdominis muscle is observed on palpation Spasm of the muscle may be due to extension of the inflammatory condition into the fibers of muscle The polymorphonuclear leukocytes range from 9 000 to 20 000 per cubic millimeter of blood after the symptoms are well established

The diagnosis of diaphragmatitis is suggested by the history and immobility of the diaphragm determined by physical signs and by roentgenoscopic examination with pain over the apex of the shoulder

Diaphragmatitis must be differentiated from spontaneous pneumothorax artificial or spontaneous pneumoperitoneum acute conditions of the abdomen which extend to the diaphragm or the transversus abdominis muscle intercostal herpes zoster pleurodynia acute spasm of the diaphragm irritation of the phrenic nerve in the mediastinum basal pneumonitis scalenus syndrome and cervical ribs

There is no specific treatment for this condition

Epidemic Hiccough (Devil s Grippe) Epidemic hiccough has been observed in rural communities The consensus is that the disease is due to an infection of viral origin The causative organism is unknown The infection apparently spreads through families by contact

The onset is sudden with epigastric and thoracic pain and increased respiratory rate *Respiration may be difficult and in some cases the rate is as high as 60 per minute* From the onset there is often a fever with the temperature from 101 to 105 F (38 3 to 40 5 C) Profuse perspiration is the rule There may be nausea and vomiting in the early stages of the disease After about 4 hours of severe paroxysms of pain and dyspnea the symptoms subside In no case do severe symptoms persist more than 24 hours without relief After 3 or 4 days from the onset *there may be* diarrhea

Examination of those who have paroxysms of abdominal pain may reveal muscle guarding and splinting In all others the presence of hiccough is the only positive finding

The diagnosis in a sporadic instance of the disease cannot be made Diagnosis is established during an epidemic mainly by the short duration of the hiccough fever and the presence of other cases in the community

The differentiation of hiccough from diaphragmatic tic is made on the rate of diaphragmatic activity *Hiccough may occur at rates of from 10 to 20 times per minute* In the diaphragmatic tic or spasm which follows encephalitis the contractures occur from 60 to 200 times per minute In diaphragmatic tic roentgenoscopic inspection of the diaphragm reveals these contractions with no other movement On forced respiration however the diaphragm is seen to descend and ascend while the small rapid contractions continue In some cases temporary or *permanent* interruption of the phrenic nerve has been necessary

In ordinary hiccough no effective treatment is known which deserves mention Inhalation of oxygen and carbon dioxide if administered by the use of a mask *prevents the patient from uttering audible complaints*

Trichinella Spiralis *Trichinella* a genus of the nematode parasites of the family Trichinellidae causes the disease trichinosis, and may cause a primary diaphragmatitis The organisms enter the diaphragm as is true of other muscles of the body and may produce serious myositis Indeed the involvement together with that of the intercostal muscles may become so extensive *as to cause* death (see Intestinal Parasites, Chapter 13)

bowel the mortality is high immediately following the accident After the acute injury has improved surgical operation may not be immediately urgent

DISEASES OF THE DIAPHRAGM SECONDARY TO DISTURBANCES OF INNERVATION OR PSYCHIC CONTROL

Paralysis of the Diaphragm Unilateral or bilateral paralysis of the diaphragm may be caused by hemorrhages poliomyelitis and tumors involving the spinal cord or the phrenic nerve Injuries or operations on the neck may crush or disrupt the phrenic nerve and thus result in temporary or permanent diaphragmatic paralysis on the same side Injury of the brachial plexus may cause paralysis of the *diaphragm A tuberculous process or malignant lymphoma adjacent to the phrenic* nerve as it courses through the thorax may involve it so as to result in paralysis of the diaphragm

When the entire diaphragm is paralyzed excitement or exertion results in marked dyspnea In unilateral paralysis symptoms are meager or absent In those who have extensive adhesions in the upper part of the abdomen severe motor symptoms referable to the intestines may follow interruption of the phrenic nerve and subsequent diaphragmatic paralysis may develop particularly when the inter ruption is of the left phrenic nerve

On examination in some instances visible paradoxical movements of the diaphragm are observed When one side of the diaphragm moves downward while the other side moves upward on inspiration paralysis is suspected

In most instances the diagnosis is made by roentgenoscopic examination

There is no treatment capable of restoring the function of the paralyzed diaphragm

Spasm of the Diaphragm (Hiccough Hiccup Singultus) Hiccough is a sudden contraction of the diaphragm If the mouth is not kept closed during the singultus a sound is produced by the quick closure of the glottis Hiccough is often produced by the swallowing of dry food without sufficient drink It is a common postoperative complication perhaps from an irritation within the abdomen It is an ominous symptom in general peritonitis Pleuritis or pericarditis may cause hiccough Involvement of the phrenic nerve anywhere along its course is a common cause of hiccough Tumors in the medulla encephalitis injuries to the phrenic nerve from wounds and trauma pressure from tumors in the neck or thorax disease of the meninges or vertebrae in the region of the third and fourth thoracic roots periph eral neuritis lead poisoning involving the phrenic nerve and inflammation of the terminal fibers within the diaphragm may result in hiccough Epidemic hiccough is thought to be associated with encephalitis Sometimes hiccough is a hysterical manifestation

The onset is sudden regardless of the cause There may be only a few con tractions of the diaphragm after which the condition completely disappears When hiccough persists pain and soreness may be experienced over the attachments of the diaphragm to the thoracic wall and of the muscles of the thorax and abdomen

Hiccough usually is a simple and harmless condition However if it persists it may lead to exhaustion and to death When the diaphragm becomes weak from prolonged hiccough the bases of the lungs do not fill well with air on inspiration and congestion results The dangers of bronchitis and pneumonia are then obvious When prolonged hiccough occurs in persons suffering from terminal diseases it often indicates that death is near

The diagnosis of hiccough is obvious A diagnosis of the cause of the symptom hiccough is often impossible The usual types clear spontaneously

Diaphragmatic Flutter Irregular clonic spasm of the diaphragm or hiccough is a common disorder in health and its significance in disease is variable Tonic spasm of the diaphragm is seen in chronic form in emphysematous patients and

Any abdominal suppurative process or abdominal operation followed by an unsatisfactory postoperative course which is characterized by chills fever pleural pain and effusion and unilateral elevation of the diaphragm arouses suspicion of the possibility of a subphrenic abscess

The most dependable roentgenologic evidence indicative of subphrenic abscess are evidences of pleural effusion and elevated diaphragm These roentgenologic findings are not conclusive but *in correlation with the history and the other findings* they are strongly suggestive of subphrenic abscess In some cases the roentgenologic findings are negative Early diagnosis of subphrenic abscess is usually dependent on a suspicion of its presence Once the diagnosis is suspected the institution of proper treatment depends on localizing the lesion as accurately as possible to determine the most satisfactory approach for surgical exploration

DISEASES OF THE DIAPHRAGM DUE TO TRAUMA

Traumatic Eventration of the Diaphragm Traumatic eventration of the diaphragm is designated also traumatic hernia false hernia and hernia spuria

Eventration of the diaphragm results from a tear in the diaphragm Such tears often are sustained from blows to the lower part of the thorax or the upper part of the abdomen accidents which result in crushing of the thorax gunshot or stab wounds or extreme straining In some cases of injury only the muscle is affected and the peritoneal and pleural coverings of the diaphragm remain intact Thus when the abdominal contents enter the thorax they carry with them the sac and may be considered as true hernias In other cases the rupture is complete whereupon the abdominal contents pass into the thorax without a sac The size of the tear determines the degree and severity of the hernia A small rent may become enlarged through pressure on the abdomen or through extreme contraction of the abdominal muscles as in straining As in congenital diaphragmatic hernia eventration occurs most frequently on the left side However both right sided and left sided rents in the diaphragm occur When a tear is situated on the right side a part of the liver itself may extend through the opening

Immediate death may result from the diaphragmatic eventration In the usual patient however the initial symptoms have commenced with nausea vomiting pain dyspnea and hiccough As the patient recovers from the acute illness he has pain in the thorax often simulating angina pectoris gallbladder colic or vague chronic digestive disorders The pain belching and feeling of fullness in the thorax are worse after meals and particularly on retiring at night when the stomach is full

On examination and on inspection soon after the accident if a large volume of the abdominal contents has entered the thorax the abdomen appears flat and there may be evidence of recent trauma such as a puncture wound or crushing injury Movements of the thoracic wall on respiration are markedly restricted

Palpation discloses that the movements of the thoracic wall are markedly decreased or absent on the involved side Tactile fremitus cannot be elicited and the heart may be displaced toward the contralateral side The percussion note depends on which part of the abdominal contents has entered the thorax If the spleen has entered or if blood has accumulated a flat note is elicited If the stomach and intestines are present a tympanitic note may be heard On auscultation because of the collapsed or compressed lung breath sounds are absent and the sounds due to air and fluid in the stomach and intestines usually heard over the abdomen may now be elicited over the thorax After recovery from the immediate effects of the accident examination by physical means may be negative

Roentgenologic examination after ingestion of a barium meal reveals evidence of parts of the *gastrointestinal organs in the thorax*

Immediate surgical operation may be indicated for the acute injury Pleurisy and peritonitis are always possible complications Because of shock and strangulated

increase in voice sounds Abscesses in the posterior mediastinum sometimes rupture into the trachea or bronchi the esophagus or pleural cavity Occasionally they appear in a posterior triangle of the neck Those which extend into the retroperitoneal space may manifest themselves in the groin

Abscesses in the posterior mediastinum may disappear soon after drainage is established through rupture into the esophagus the trachea or a bronchus

The diagnosis is seldom made until rupture occurs However if the condition is suspected the ingestion of barium sulfate by the patient enables visualization of the esophagus displaced ventrally Often there is evidence of obstruction due to pressure Abscesses in other mediastinal parts if they should occur are not diagnosable

MEDIASTINAL EMPHYSEMA

Air in the mediastinum may follow severe coughing crying lifting straining at stool thoracic injuries overdistention of the lungs surgical procedures on the lungs and pleura the use of positive pressure anesthesia spontaneous interstitial pulmonary emphysema pulmonary diseases such as bronchopneumonia influenza asthma and tuberculosis and a ruptured abdominal viscus

On reaching the mediastinum air travels along courses of least resistance toward the cervical occasionally toward the abdominal regions In the neck air localizes in the subcutaneous tissues where it is detected by means of the crepitations elicited on palpation The air reaching the retroperitoneal space may rupture through the peritoneum and result in pneumoperitoneum or it may course downward through the inguinal rings causing subcutaneous emphysema of the thighs and abdominal wall Air in the peritoneal cavity when there is no rupture of an abdominal viscus usually arrives there from the lungs

Symptoms Often the escape of air into the retroperitoneal space and into the region of the neck is accomplished with but mild discomfort However if air reaches the mediastinum under high pressure as for instance from a high pressure pneumothorax the pain is excruciating The pain may extend to the left shoulder and the left arm There is a sense of pressure beneath the sternum Swallowing and turning the head are painful There are severe dyspnea and cyanosis When the intramediastinal pressure approaches zero the return flow of blood to the auricles is lessened through compression of the systemic and pulmonary veins

Examination There is often swelling of the cervical region when the subcutaneous emphysema is marked The presence of swelling over the abdomen the inguinal region and the thighs may signify subcutaneous emphysema in these regions

A subcutaneous emphysema is characterized by crepitations on palpation of the subcutaneous tissue When considerable air has accumulated in the mediastinum the percussion note over the sternum becomes hyperresonant On auscultation crackling popping sounds are heard over the sternum and over the surface of the neck and along its borders which are increased when the heart is in systole

Diagnosis The physical findings are diagnostic In instances in which diagnostic uncertainties prevail lateral roentgenograms may be very helpful

Usually the prognosis is excellent In most cases of mediastinal emphysema treatment is not required

In instances of acute mediastinal emphysema the syndrome of acute coronary occlusion may be simulated A short period of observation plus electrocardiographic studies will identify the condition present

TUMORS OF THE MEDIASTINUM

Infections may reach the mediastinal lymph nodes from the neck the abdomen the lungs and pleura and the surface of the thorax and cause them to enlarge The tubercle bacillus is the chief cause of the enlargement but other agents also such

cannot be made by any means of examination It is suspected if the infection extends to the neck or downward into the retroperitoneal tissues or into the hili of the lungs

Acute Mediastinal Abscess The etiology of acute mediastinal abscesses is the same as that of acute mediastinitis Abscesses in the anterior mediastinum are caused by extensions of infection from lymph nodes and by erysipelas and occur after operations on the lungs the larynx the neck and the sternum Occasionally a mediastinal abscess is a complication of disease in the abdomen for instance a gall bladder infection and particularly perforation of the gallbladder

There is moderate fever and pain and throbbing are present beneath the sternum The pain is accentuated by *movements of the trachea* as *by coughing*

On examination often the veins of the neck are distended There are swelling and edema of the neck and thoracic wall Increased percussion dullness is often elicited over the manubrium Leukocytosis is present

The diagnosis may be obvious from the foregoing physical findings in association with roentgenologic evidence of a mass displacing the trachea At times if there should be only miliary abscess the diagnosis may not even be suspected clinically

Chronic Mediastinitis Chronic mediastinitis is caused by pyogenic infections rheumatic fever syphilis and tuberculosis Unusual causes of chronic mediastinitis are actinomycosis or streptotricosis pneumoconiosis and chronic paragonimiasis The following general statements can be made in regard to chronic mediastinitis (1) The chronic lesions produce fibrosis which may constrict the innominate veins or the superior vena cava Edema of the face neck and upper extremities and the appearance of collateral circulation ensues (2) If the infection extends to and involves the pericardium there are edema of the feet and legs ascites dyspnea cyanosis and cough (3) If collateral circulation is established there are enlarged superficial veins over the thorax and abdomen As the scarring tightens stenosis of the trachea or bronchi ensues which may be evident symptomatically and on roentgenologic examination Paralysis of the recurrent laryngeal nerves is manifested by changes or loss of voice

The roentgenologic examination confirms the symptoms and the physical findings by revealing a dense shadow which protrudes laterally slightly beyond the costosternal borders Such a roentgenologic finding is diagnostic unless accompanied by a history and the presence of a known disease which could cause a mediastinal fibrosis or tumor

Chronic Mediastinal Abscess ***Chronic Abscess of the Anterior Mediastinum*** Chronic abscesses in the anterior mediastinum may be caused by tuberculosis syphilis actinomycosis and infected mediastinal dermoid cyst If there should be symptoms they result from the primary disease for these abscesses are usually without manifestations of their own

Percussion directly over the abscess yields dullness and often tenderness This is the only finding on physical examination suggestive of abscess in the anterior mediastinum The findings on percussion when taken in conjunction with the history and the results of roentgenologic examination may be diagnostically conclusive

Chronic Abscess of the Posterior Mediastinum An abscess of the posterior mediastinum may be caused by perforation of the esophagus or by extension of infection from osteomyelitis of a vertebra or from infection in the retropharynx the lung pleura or abdominal cavity Occasionally an abscess may arise in situ from diseased mediastinal lymph nodes

The first symptoms are those of pressure There are pain dysphagia cough and dyspnea The pain is increased by swallowing or coughing It is situated between the scapulae and often extends around toward the front of the thorax The abscess may rupture into a bronchus with expectoration of pus Esophageal rupture creates a serious condition

On physical examination there may be dullness over the spinal column with

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as infectious mononucleosis syphilis Hodgkin s disease sarcoma sarcoidosis and carcinoma cause enlargement of these nodes

In children a lobe of the thymus protrudes laterally so as to cast a roentgeno logic shadow which may resemble that present in disease of the upper pulmonary lobe Tumors of the thymus or persistent thymus may be revealed by roentgeno scopic examination

Likewise the thyroid gland may extend substernally into the thorax and result in pressure on the trachea esophagus internal jugular vein carotid artery and the recurrent laryngeal and cervical sympathetic nerves

Cysts of the mediastinum may be congenital inflammatory or parasitic in origin The common tumors of the mediastinum are congenital namely dermoid or tera toma cystic lymphangioma pericardial celomic cyst bronchial cyst gastric cyst and enteric cyst

Benign tumors as well as malignant growths often result in pressure symptoms

SYMPTOMS Tumors of the mediastinal structures are manifested by symptoms referable to the circulatory system the respiratory system the mediastinal nerves and the digestive system These symptoms are very much the same whether pro duced by a benign or a malignant growth since they originate largely from pressure of the enlarging tumor

Circulatory Symptoms When obstruction of the superior vena cava is present the patient on arising after stooping or flexing the body for a minute or two has cyanosis of the face neck and upper part of the thorax The superficial veins of these regions become more dilated and engorged than those of a normal person who per forms a similar act The engorgement of the face in cases of mediastinal pressure will remain for some time in contrast to the rapidity with which this flushing will dis appear in the normal person In instances of marked obstruction the face may be a bluish black markedly bloated and the eyes may bulge as is seen in strangulation Venous obstruction may produce cerebral congestion resulting in dizziness tinnitus headache and even loss of consciousness Instances of unilateral venous obstruction often can be demonstrated by having the patient raise both arms to a horizontal position above the level of the upper limits of the heart chambers If unilateral obstruction should be present the superficial veins of one arm will become distended and remain so while the arm is maintained in the horizontal position

Respiratory Symptoms A patient who has mediastinal pressure producing res piratory symptoms has dyspnea and cough The dyspnea at first appears on exertion As the pressure increases it is paroxysmal or continuous

In the beginning the cough is dry Later when sputum appears it is mucoid and finally it becomes mucopurulent and hemoptysis is common The cough and expectoration result from pressure and obstruction of the air passages and thus from an obstructive bronchitis

Respiratory symptoms may be due to traction or pressure on the trachea or large bronchi or to circulatory obstruction Respiratory stridor and wheezing result from pressure causing narrowing of the trachea or bronchi Occasionally these symptoms are associated with involvement of the recurrent laryngeal nerve

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Symptoms of Involvement of Mediastinal Nerves A patient who has a medi astinal tumor which is irritating the recurrent laryngeal nerves has hoarseness or complete aphonia There may be the brassy type of cough formerly considered characteristic of an aneurysm of the aorta

As the result of pressure on the sympathetic nerves constriction of the pupil on the affected side is frequently an early manifestation Pressure on a sympathetic nerve may be manifested by unilateral sweating and flushing of the face Vagus

pressure resulting in a slow pulse and hiccough from phrenic nerve involvement is rare but does occur

A dull aching pain is usually an early symptom Paroxysms of intense pain or a prolonged period of excruciating type of pain may be witnessed There is a sense of oppression or substernal tightness Pressure on the intercostal nerves may result in neuralgia or neuritis and herpes zoster Pleuritic pain is frequent Erosion of the vertebrae or the sternum produces the boring pain which occurs so often at night in all bone diseases

Examination On examination there may be observed exophthalmos and dilatation contraction or inequality of the pupils Edema cyanosis or unilateral flushing and perspiration of the face may be apparent There is dyspnea on exertion early in the disease If dyspnea should be well established on slight exertion or while at rest orthopnea is present

A slight fullness of the intercostal spaces near the sternum or a bulging of the sternum in the midline is usually present in dyspneic patients A nonexpansile pulsation of the bulging region is likewise often observed The tumor may be visible in the sternal notch or above the sternoclavicular articulation If the mediastinal involvement results from a tumor which arose in the neck this fact is obvious

Evidences of interference with the circulatory organs are manifested late in the disease There is usually unilateral dilatation of the veins of the thorax neck and occasionally the face The jugular veins are prominent and the veins between the clavicle and the third or the fourth rib anteriorly are distended and tortuous Occasionally the distended and tortuous veins can be seen on the back below the diaphragm Local cyanosis and edema corresponding in area with the venous dilatation are constantly present

The neck the sternal notch and the axilla are palpated to determine the presence of tumors or enlarged lymph nodes Enlarged nodes may be felt in the axilla in the supraclavicular region or along the lower border of the pectoral muscle Palpation may reveal deviation and fixation of the larynx and trachea A tracheal tug is more frequently associated with an aortic aneurysm but it may be present in other mediastinal tumors There is often a decreased expansion of the thorax The pulse may be slow from vagal irritation or fast from a weak heart Inequality of the two radial pulses from unilateral interference with the arterial circulation is often present

Submanubrial dullness may be elicited on percussion This dullness rarely can be demonstrated however unless it extends beyond the sternal edge The area of dullness is often irregular in outline and extensive In some posterior tumors percussion over the thoracic spines may demonstrate an abnormal flatness to percussion unless the tumor is large however there will be no definite dullness on percussion In large tumors there is decreased or absent vocal fremitus over the area of percussion dullness

On auscultation in some instances of large tumors there is an increased transmission of tracheal respiration in the dull area over the manubrium and absent or diminished transmission of voice or breath sounds over other areas If there should be pressure on a main bronchus respiratory sounds on one whole side of the thorax may be suppressed or absent Medium coarse piping or musical rales may be heard in one or both lungs as a result of pressure of bronchitis

Diagnosis The presence of manifestations of mediastinal pressure which affects the mediastinal circulation respiration or nerves singly or in combination is clinically unmistakable The nature of the tumor as to whether it is benign malignant or of vascular origin (aneurysm) can often be determined by the roentgenologist

Teratomas and dermoid cysts occur behind the sternum rarely in the posterior mediastinum in patients less than 30 years of age Difficulty in diagnosis of these follows perforation into a bronchus with the ensuing infection

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Symptoms of Involvement of Mediastinal Nerves A patient who has a medi astinal tumor which is irritating the recurrent laryngeal nerves has hoarseness or complete aphonia There may be the brassy type of cough formerly considered characteristic of an aneurysm of the aorta

As the result of pressure on the sympathetic nerves constriction of the pupil on the affected side is frequently an early manifestation Pressure on a sympathetic nerve may be manifested by unilateral sweating and flushing of the face Vagus

Inspection does not reveal abnormalities On first palpation and percussion there are no abnormal signs By the time the sputum begins to be profuse and to collect in the bronchial tubes vibrations produced by it on breathing may be felt on the surface of the thoracic wall On auscultation there are sibilant sonorous bronchial rales Later coarse moist and bubbling rales are heard Coughing may result in a temporary disappearance of rales The breath sounds may be masked by the numerous rales throughout the thorax The physical signs in acute bronchitis are usually elicited equally well over both sides of the thorax When the physical signs are encountered over only a part of one lung the condition therein is a more serious condition than acute bronchitis

Diagnosis is established by physical examination Acute bronchitis may be a serious disease in infants and elderly persons especially in those who have emphysema asthma or heart disease In otherwise normal persons recovery is the rule Occasionally an acute bronchitis progresses and pneumonia develops

Acute Bronchiolitis (Capillary Bronchitis) An acute inflammation mainly of the walls of the bronchioles is caused by bacteria and probably viruses The first symptoms are often those of an acute respiratory infection The symptoms become more severe and soon dyspnea and then cyanosis develop The temperature rises to 103 or 104 F (39 4 or 40 C) The cough is distressing Moderate prostration ensues

The patient appears to be seriously ill There are cyanosis profuse perspiration shallow and rapid breathing Findings on palpation and percussion are normal throughout the lungs Auscultation reveals early in the course of the disease sibilant rales and later fine moist rales throughout the lungs

Cultures of the sputum reveal a preponderance either of the influenza bacillus or of the hemolytic streptococcus Occasionally staphylococci are present in abundance In some instances there is leukopenia and in others leukocytosis is present

The diagnosis of acute bronchiolitis cannot be made from physical examination The results of roentgenologic examination likewise are indefinite This condition simulates a diffuse bronchial pneumonia If the course is favorable the diagnosis may not be made otherwise the diagnosis awaits necropsy

Oxygen chemotherapy or antibiotics are desirable

Chronic Bronchitis Chronic bronchitis in adults occurs in those debilitated by tuberculosis emphysema circulatory insufficiency bronchiectasis allergic conditions infection of the paranasal sinuses pneumoconiosis fungous infections fusospirochetal infections foreign bodies dust and benign or malignant tumors The aged are prone to have this disease

The cough is constantly present and tiresome Soon after meals the cough may cause vomiting and thus adds to the weakness and often loss of weight The cough and expectoration are always worse on breathing of cold damp air The muscles of the lower thoracic wall and particularly those of the upper abdominal wall may become sore and tender to a degree that an intra abdominal disease may be suspected

The eyes are often reddened and the lids may be hemorrhagic from the cough An emphysematous shaped chest is present The lungs are hyperresonant on percussion and there are musical rales on auscultation These may clear on deep breathing Medium and fine rales may be elicited over the bases which do not clear on deep breathing The breath sounds may be somewhat diminished and the expiratory phase of respiration is longer and lower pitched than usual On palpation of the upper part of the abdomen there may be diffuse soreness and tenderness of the muscles Palpation and percussion often reveal evidence of primary disease of the circulatory apparatus

The diagnosis is established by a process of elimination of primary disease such as tuberculosis heart disease malignant disease fungous infections and pressure on the trachea or bronchi

Malignant tumors of the thymus may be suspected if there should be the accompanying symptoms of myasthenia gravis Lymphosarcoma (Hodgkin s type) is usually manifested by involvement of the lymph nodes of the neck before there is manifest mediastinal pressure

Aneurysms can usually be determined by roentgenoscopic examination

Invasion of the mediastinum by malignant neoplasms from elsewhere in the body occurs by direct extension or by lymphogenous or hematogenous spread Direct extension to the mediastinum occurs in carcinoma of the trachea bronchi esophagus and breast Carcinoma of the breast may also metastasize by the blood stream or the lymph stream Lymphatic and blood stream metastasis to the mediastinum is extremely rare The symptoms and physical findings are evidence of pressure on one or more of the mediastinal contents Diagnosis usually depends on recognition of the primary tumor

Except for secondary invasion of mediastinal structures by sarcomatous growths sarcoma is rare Metastatic involvement of the mediastinum from axillary melano sarcoma and bone sarcoma has been reported Besides the signs and symptoms of the primary growth those of pressure or invasion of mediastinal structures may be found

DISEASES OF THE BRONCHI

The Bronchial System The *right main bronchus* is larger than the left and is more nearly a continuation of the trachea The right main bronchus divides into the right upper the middle the dorsal and lower branches

The *right upper lobe bronchus* divides into three segmental branches (1) right anterolateral segment (2) right posterolateral segment and (3) right apical segment

The *right middle lobe bronchus* divides into two major branches one of which supplies the right anterior middle segment and the other supplies the right lateral middle segment of the lung

The *dorsal bronchus* divides into the anterior posterior and middle bronchi which supply lower lobes of the lung

The *lower branch* divides twice into the mid posterior and anterior basic branches

The *left main bronchus* is immediately more branching in character than the right *main bronchus The first division of the left main bronchus divides into* the left upper lobe bronchus the anterolateral the lateral mid anterior mid anterior basic mid basic posterior basic and dorsal bronchi

The Bronchi and Lungs as Originators of Pain Clinically according to Jackson pain is almost never noted during bronchoscopic manipulations even in dealing with sharp pointed foreign objects which inevitably inflict some trauma Occasionally a patient with a bronchial foreign body is able to localize it correctly as to side Real pain is extremely rare in these cases unless there is pleural involvement Pain does appear to be felt more in the bronchi than in the trachea but possibly this is due to the proximity to the pleura

Acute Bronchitis Acute bronchitis is usually a part of a catarrhal inflammation of the bronchi trachea and larynx It frequently begins as an acute infection of the respiratory tract

Infections by viruses and by bacteria often aided in the beginning by mechanical irritants certain dusts fumes gases and inhaled substances to which the tissues are allergic cause what is clinically recognized as acute bronchitis

The first symptom of acute bronchitis may be cough A burning or raw sensation in the throat and beneath the sternum may be distressing From 1 to 2 days after the onset the sputum may become profuse Quantities of yellowish greenish stringy mucous material sometimes with a fetid odor and often blood streaked may be expectorated In some instances the onset may be with fever (temperature 100 to 101 F or 37 8 to 38 3 C) There is generalized aching particularly severe over the lumbar region and down the legs Except in those who have asthma and chronic conditions of the bronchial tree the symptoms pass in 3 to 5 days

The age and sex of the patients fail to show marked differences in the types of the disease Blood spitting is commoner with saccular dilatation than with other types In Chapman and Wiggins series of cases blood spitting had occurred in 42 per cent for more than 2 years About one half estimated the amount at 1 ounce (30 ml) or less in 24 hours About 40 per cent were unaware of having had pneumonia The incidence of pneumonia in saccular bronchiectasis was greater than in the cylindric type Influenza if present was unknown to 27 per cent There was no significant difference in its incidence among the three types Nasal disease was a complaint of 35 per cent Its incidence was about the same in the three types Physical signs were absent more often in cylindric bronchiectasis than in the other types The only patients known to have asthma were in the cylindric group These data on the distribution of the disease differ in that there was a relatively higher incidence in the right lower lobe as compared with the left This is unexpected in view of the fact that congenital bronchiectasis should involve the left lower lobe more commonly It appears that distinction between cylindric and saccular bronchiectasis is based on sound statistical grounds This is not the case with other morphologic types

In advanced disease extensive fibrosis emphysema and cavities are present Occasionally an abscess may appear in a part of the lung not involved by bronchiectasis as the result of aspiration of infected material from the involved bronchi

In the so called dry type of bronchiectasis the parenchymal changes are slight but in some patients the fibrosis continues to develop until it extends throughout the lung or lungs As the fibrosis progresses compensatory emphysema obscures the physical signs of the disease

There are both congenital and acquired forms of bronchiectasis Regarding the congenital form of bronchiectasis Olsen stated that the condition in the lungs is manifested in almost half of the cases before the age of 10 years and that it follows an attack of influenza measles pertussis or pneumonia Often there is an associated dextrocardia

Acquired bronchiectasis develops after streptococcal bronchopneumonia either in childhood or in adulthood First the changes involve the bronchial mucosa the submucosa and the pulmonary parenchyma Progression of these changes results from recurring bronchopneumonia The chronic stasis and infection distal to a bronchial stenosis or obstruction from any cause sequentially induce development of bronchopneumonia pulmonary abscess and later indurative pneumonia and bronchiectasis

Symptoms Small regions of bronchiectasis are often symptomless but such small regions may be the point of origin of severe hemorrhages In those who have symptoms paroxysms of coughing when the patient changes position often soon after getting out of bed in the morning are common The cough may or may not bring up large quantities of sputum The paroxysms of cough may last from a few minutes to an hour or more

Pulmonary hemorrhage is a common complaint of those who have bronchiectasis Vinson found that in 100 cases of chronic bronchiectasis there were histories of pulmonary hemorrhage in 49 According to Vinson these pulmonary hemorrhages are not predictable on the basis of degrees of involvement of the lung tissues or of prodromal symptoms

In many instances the patient often has observed that on bending forward while coughing a large amount of sputum is obtained

Examination Inspection reveals widening of the intercostal spaces retraction and lagging of the thoracic wall and perhaps the absence of the phrenic wave signs (Litten s sign) Clubbing of the fingers and the toes often appears soon after the onset of the symptoms The clubbing may involve only the soft tissues or may be associated with hypertrophic pulmonary osteoarthropathy Dyspnea is present when there are advanced pulmonary fibrosis and emphysema

On palpation tactile fremitus may be increased or normal depending on the amount of pulmonary fibrosis and its proximity to the periphery of the lungs

Fibrinous Bronchitis This form of chronic bronchitis is called also pseudo membranous, membranous croupous mucinous and plastic bronchitis

The etiology of fibrinous bronchitis is not known The condition in addition to other symptoms of bronchitis is characterized by the expectoration of casts of fibrinous mucous material The casts have the shape of the bronchial ramification in which they were formed

In the acute stage of the disease cough chills and fever are present which are followed by pain and dyspnea and expectoration of the casts In the chronic stage the symptoms are mild but prolonged Hemoptysis is common after expectoration *of the casts*

On examination the findings are those of chronic bronchitis In diagnosis the presence of the casts in the sputum is the only distinguishing feature of this form of chronic bronchitis from all others

Tracheobronchial Tuberculosis Tracheobronchial tuberculosis is frequent in primary pulmonary tuberculosis of children and adolescents and in the re infection type of pulmonary tuberculosis in adults

The bronchial mucosa is frequently the seat of both tuberculous and non tuberculous lesions The bronchi are either compressed by the enlarged lymph nodes or retracted by sclerosis of the pulmonary lesions The evolution of tuberculous bronchial lesions runs parallel with the evolution of the tuberculous process in the involved lobe Acute exudative bronchial tuberculosis appears in some cases years after the regression of the pulmonary tuberculosis and its calcification

The symptoms are often those of an asthmatic or chronic bronchitis The diag nosis is established by demonstration of tubercle bacilli and by bronchoscopic and roentgenologic examination followed by histologic and cultural examinations of biopsy tissue

Broncholithiasis (Pneumoliths or Lung Stones) Broncholiths are composed largely of calcium Lung stones may be present in inspissated pus in regions of tuberculous caseation in lymph nodes and lungs around aspirated foreign bodies

Broncholiths may develop within the bronchi or may originate outside the bronchi *with subsequent perforation into the air passages In the majority of cases* broncholiths are due to perforation of calcified tuberculous lymph nodes into the bronchus The changes found in the presence of broncholiths may simulate carci noma of the lung chronic pulmonary abscess bronchiectasis with atelectasis and chronic pneumonitis as well as mycotic disease Body section roentgenography is of diagnostic aid in bronchopulmonary lithiasis

The clinical symptoms may vary from complete absence of signs or symptoms *to those of a severe or critical illness Practically every pathologic* condition which may develop in the thorax may be simulated Bronchopulmonary lithiasis should be included in differential diagnosis of bronchial obstruction pulmonary suppura tion and hemorrhage associated with paroxysmal attacks of cough when the thoracic roentgenogram shows calcific shadows in the region of consolidated *portions*

The diagnosis is established on bronchoscopic inspection by finding the injury of the bronchial wall through which the stone has passed The diagnosis is not fre quently made The patient expectorates a stone followed by a small amount of blood and the illness is over without the physician knowing anything about it

Bronchiectasis Bronchiectasis is characterized by irregular sacculations of the walls and variations in caliber of the lumina of the smaller bronchi These structural *aberrations of the bronchi are often segmental in distribution* affecting a small part of one lobe of either lung in the base or the disease may affect bronchi throughout the greater part of both lungs

The types of bronchial dilatations by roentgenographic study are the cylindric saccular varicose fusiform and bronchiolectatic *with the cylindric* saccular and varicose forms being the common types

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Fremitus is decreased if the pleura is thickened In the presence of emphysema of the lungs there is a decrease in tactile fremitus

On percussion changes are present in those who have a thickened pleura and emphysematous regions

The auscultatory findings are often within normal limits Occasionally there are fine to coarse and intermittent rales, depending on whether or not the contents of the sacculations of the bronchi have been expectorated The breath sounds may be bronchial or cavernous when there are large empty bronchiectatic cavities

On endoscopic examination there may be discovered a foreign body or some other obstruction the removal of which if feasible facilitates treatment or cures the patient

With the head and shoulders nearly to the floor forced coughing will often be followed by the expectoration of a large amount of sputum Within a 24 hour period a volume of sputum equaling 2 pints (1 liter) or more may be expectorated The sputum is often blood streaked and in it are greenish yellow masses ½ inch to 1 inch (1 27 to 2 5 cm) in diameter (Traube s or Dittrich s plugs) When allowed to stand the sputum separates into three layers The bottom layer is opaque with pus cellular debris bacteria and occasionally elastic fibers the middle layer appears somewhat clear and watery and the top layer is frothy The sputum is examined for tubercle bacilli since tuberculosis may coexist with or be the cause of bronchiectasis

DIAGNOSIS The diagnosis is suspected from the history and is confirmed on the examination Usually the pulmonary involvement is detected on roentgenologic examination An endoscopic examination is often invaluable in detection of a localized bronchial stenosis or of a foreign body obstructing a bronchus

Postural drainage should be instituted at once The surgical removal of the involved lobe or even the entire lung when necessary is satisfactory if the patient survives the operation and the patient is not left with an incapacitating postoperative thoracic pain

Bronchostenosis Bronchostenosis may develop from pressure extrinsic to a bronchus or from inflammation or growths from within its walls Conditions which cause bronchostenosis also cause bronchial occlusion Prickman and Moersch examined endoscopically 140 patients suffering from asthma in 60 of whom definite stenosis was found in one or more bronchi

A significant and prominent symptom is severe persistent and sometimes paroxysmal cough often of long duration and dating from an acute respiratory infection At times it may be impossible for the patient to cough up the sputum Then at times the cough loosens and there is profuse mucopurulent sputum streaked with blood This increased amount of sputum represents some of the secretions which have been retained by the stenosis Bronchostenosis may result in pneumonitis and atelectasis and if not relieved bronchiectasis may ensue

The physical signs are those of pneumonitis and atelectasis Often a wheeze is elicited and occasionally crepitant rales are heard

A presumptive diagnosis of bronchostenosis is often made from the history and physical findings The exact diagnosis is made only by the bronchoscopist or the roentgenologist

Bronchial Perforation Acute or chronic pyogenic abscesses chronically inflamed lymph nodes containing lung stones and lesions of chronic pulmonary tuberculosis may perforate ramifications of bronchi and discharge their contents into the bronchial system Materials thus discharged are distributed peripherally and in the case of tuberculous material acute pulmonary tuberculosis may ensue

The symptoms are those of the prevailing disease

Results of examination often are negative In some instances tubercle bacilli may be found in the sputum or gastric washings although the lungs appear entirely clear on examination

The diagnosis is made by a capable bronchoscopist

Bronchial Fistulas Bronchial fistulas are often the sequence of perforations of the viscera of the abdominal cavity After perforation of abdominal viscera a sub phrenic abscess forms with subsequent perforation of the diaphragm and into a bronchus In the course of empyema or subphrenic abscess bronchocolic fistula may develop

The symptoms are obscured by those of subphrenic abscess

The diagnosis is often difficult however Escherichia coli found in abundance in the sputum or an abundance of pus in the feces is suggestive Bronchography may suffice if the iodized oil enters the fistulous tract and empties into the colon If this method fails a barium enema may be helpful if the barium enters the bronchial system When the fistula fails to close surgical treatment may be indicated

Bronchobiliary Fistulas Bronchobiliary fistulas develop when hepatic ab scesses penetrate the diaphragm and a communication is established with one of the larger bronchi Common causes are amebic and echinococcal infections of the liver The sequence of events in an occasional case is the formation of gallstones followed by suppurative cholangitis with perforation and formation of a small sub phrenic abscess with perforation of the diaphragm and a bronchial ramification

The diagnosis can be made when the patient expectorates bile In some cases roentgenologic examination following the administration of iodized oil reveals the fistulous tract When such fistulas do not close surgical intervention may be con sidered

Broncho esophageal fistulas are usually fatal However an occasional patient is observed who has a fistula which has slowly developed as the result of some chronic disease in the esophagus or in a bronchus who has lived for a long time In rare instances broncho-esophageal fistula has followed pneumonia and the patient may have lived for a long time on very low fluid intake to avoid coughing when liquids were swallowed

The symptoms of a broncho esophageal fistula are cough and dysphagia im mediately after swallowing food or drink unless the liquid is taken in sips Parox ysmal half vomiting and half coughing characterize this type of dysphagia

It is important to determine the general physical condition and to discover evi dence of the underlying disease There are no physical signs indicative of these fistulas

If one of these fistulas is suspected endoscopic examination of the esophagus is diagnostic

Bronchial Obstructions and Pulmonary Atelectasis Xalabarder defined atelec tasis as the absence of air in the pulmonary alveoli with intense retraction and shrinking the latter differentiating it from simple pulmonary collapse As a result of the vascular dilatation characteristic of atelectasis intra alveolar exudates from simple serous edema to hemorrhagic suffusions are produced Reversion is likely to occur but if inflammation persists a long time a progressive fibrosis prevails The mechanisms of the production of atelectasis are not always obvious for (1) there are bronchial obstructions without atelectasis (2) the existence of bronchial plugs is not constant (3) atelectasis can be produced by obstruction of a main bronchus but not of a secondary bronchus and (4) once the gaseous pressure on each side of the alveolar wall has become equal the alveolus will be relaxed to the maximal limit which its elastic structure permits

Xalabarder has expressed the belief that atelectasis is produced by pulmonary contraction and that it originates from several causes acting on a specially pre disposed organ One of these causes may be bronchial obstruction but this is not an indispensable factor Atelectasis is the abnormal exaggeration of the physiologic pulmonary tone This defective function is of importance in the genesis of other pulmonary phenomena including the active dilatation of the lung (insatiable pneu mothorax and functional emphysema) Xalabarder accepted the existence of muscu lar fibers in the alveolar walls and saw in the functioning of these muscular plexuses

under regulation by the autonomic nervous system the solution to a regional contracture in the affected pulmonary zone that is to say to atelectasis

Congenital Pulmonary Atelectasis Respiratory movements take place during the second half of intra uterine life and thus some of the intrapulmonary spaces are occupied by amniotic fluid This fluid may cause changes in the lungs which prevent full expansion of the lungs at birth A failure on the part of the lung to expand may also be due to the congenital absence of alveoli to bronchial atresia or to obstruction of bronchi

A widespread atelectasis may prevent breathing by the newborn child In the lungs of the newborn there may be regions of atelectasis which are permanent and which do not cause symptoms These regions of the lung are sealed off and leave only a strand of fibrous tissue When regions of the lung thus degenerate fully symptoms never occur

In the lungs of some adolescents and adults there is a well-established bronchiectasis which has been present since early childhood without inflammation of the lungs These regions represent the so called dry bronchiectasis which may be discovered during life or may be found at the time of postmortem examination The regions are often situated in the lower lobe of the left lung In other instances these regions may become infected and the manifestations of atelectasis ensue

Obstructive or Acquired Pulmonary Atelectasis Normally the bronchi are filled with air and the right amount of secretion The secretion from the bronchi is drained upward This upward drainage is afforded by the pumping effect of respiration aided by the upward transportation of small particulate matter by the ciliary activity of the cells lining the bronchi and finally by the expulsive force of a cough

When a mechanical obstruction develops its effects depend on its degree on whether the obstruction is complete or partial and if partial on whether it is fixed or movable

Complete bronchial obstruction is immediately followed by absorption of gases and total retraction of the involved part of the lungs There may still remain functioning pulmonary tissue in the region of the obstruction which is supplied by a bronchus tributary to the obstructed bronchus Inflammation or edema interferes with absorption of gas and retraction For instance a lobe of the lung that is distal to complete obstruction by a foreign body may show evidence of atelectasis edema and inflammatory infiltration but owing to an anastomosing communication between the alveoli of each lobe it remains partially or wholly distended These communications between the lobes of the lung are a protection against atelectasis that follows occlusion of the smaller bronchi

Pulmonary atelectasis occurs when there is compression of pulmonary tissue or an obstruction to a bronchus In either case the lung is unable to expand on inspiration There are several conditions which reduce the size of the thoracic cavity sufficiently to produce atelectasis for instance fluid or air in the pleura removal of ribs or paralysis of the diaphragm Under these circumstances the lungs are forced to adjust themselves to the new volume by retraction Lesions of the central nervous system that cause paralysis of the muscles of inspiration are followed by atelectasis

An obstruction of a bronchus with subsequent retraction of the lung after the air has been absorbed from it is a common cause of segmental atelectasis

Postoperative Atelectasis (*Postoperative Pneumonia*) During and after surgical operations because of pain and sedation the respirations become shallow the cough reflex is suppressed and there is an immobilization of the thorax in a fixed posture As the result of these abnormal conditions aspiration and stagnation of secretion from the upper part of the respiratory tract ensue and often atelectasis supervenes The condition in the lung may remain atelectatic or may become pneumonic Thus the postoperative atelectasis is often a postoperative pneumonia

Postoperative pulmonary obstruction and consequent atelectasis or pneumonia are commoner when acute respiratory infection is present at the time of operation or when the patient has been exposed to any acute respiratory infection just prior to operation

From one to four days after the operation if the patient is mentally alert there

is a feeling of tightness or constriction in the lower part of the thorax manifested as an oppression during breathing In extensive involvement there are dyspnea cyanosis cough and expectoration In all however whether there are complaints or not during the first three or four postoperative days the temperature rises to 101 F (38 3 C) or higher The pulse rate will be fast (120 or more)

In mild illness temperature and pulse rate return to normal within a week The pulse rate returns to normal before the fever subsides

In many patients the illness is prolonged and there is pneumonitis or pneumonia finally terminating in bronchiectasis or death

On examination when feasible there may be relative dullness to percussion enfeebled breath sounds and fine rales over the lower lobe or lobes Bronchial breathing is often present On occasion a pleural rub may be heard There is retraction of the intercostal spaces during inspiration The diaphragm is elevated and the apex beat of the heart may be shifted toward the involved side

The diagnosis depends on the history postoperative course physical findings and results of roentgenologic examinations It is inadvisable to examine very ill patients physically and in some cases roentgenologically

Infection Atelectasis During the course of respiratory infections degrees of atelectasis are often present The small atelectatic portions of the pulmonary tissue assume normal volume and function as soon as the infection subsides Larger regions resulting from obstruction of larger bronchi become clinically manifested Long after a bad cold has passed a cough and expectoration persist which may be indicative of an unknown atelectasis

During the course of an acute respiratory infection there is acute thoracic pain which is situated in the base of the lung beneath the scapula around to the axillary line and dyspnea which if severe is accompanied by cyanosis When these symptoms are present the occlusion is large and there is a virulent infection Under these conditions fever and sometimes chills develop

On physical examination there is elevation of the diaphragm and shifting of the mediastinum Over the atelectatic portions of the lung there are altered or absent breath sounds rales and varying degrees of consolidation

The history the physical findings and the findings on roentgenoscopic examination are diagnostic These patients should have an endoscopic examination

Alveolar and Pulmonary Adenomatosis Benign pulmonary adenomatosis is a disease with multiple involvement of the lungs by a process in which the alveoli are filled with distended columnar cells The similarity between this adenomatous process and the metastasizing alveolar cell tumor is sufficiently close to suggest that pulmonary adenomatosis is a potentially malignant tumor However extrapulmonary metastasis is unknown in adenomatosis

Pulmonary adenomatosis may have a possible relation to jagziekte an epizootic pulmonary adenomatosis of sheep which is considered to be a viral infection Like pulmonary adenomatosis metastasis does not occur in jagziekte The attempts to transmit the condition to animals from material obtained from human beings with pulmonary adenomatosis have failed

The sex incidence is two women to one man The age of the patients has ranged from 30 to 80 years

The symptoms commence with cough which varies from nonproductive and mild to productive and severe This consistent complaint is attended by fatigue weakness dyspnea wheezing and night sweats In some the manifestations are chronic coughs followed by a pneumonia of lobar distribution Because of the diffuseness of the process dyspnea can appear very early and be out of proportion to cough and expectoration

Sputum is profuse frothy tenacious and there is difficulty in expectorating it despite the fact that there is constant welling up of the material into the throat The

EXAMINATION The physical findings in carcinoma of the lung vary with the location size and rate of growth of the neoplasm Often there are no abnormalities on examination by physical means until there are coexistent pathologic change and complications secondary to the tumor The primary malignant disease may thus be obscured

In well advanced cancer of the lung the thoracic movement may be limited or absent The limitation of thoracic movement in the beginning is confined to the affected side but later both sides are affected If a sufficient volume of effusion is present the intercostal spaces bulge As the pressure increases in the thorax the veins of the neck and thoracic wall are distended and edema of the face neck or arms is present On palpation enlarged lymph nodes may be felt in the lower triangles of the neck and in the axilla

When the disease is advanced tactile fremitus is decreased or absent over the diseased side and often is increased on the normal side The cardiac impulse may be displaced toward the side of the pulmonary lesion ✓

There are no changes in the percussion sounds until a considerable portion of the pulmonary tissue is hardened by the tumor Then the percussion sound is dull or flat Dullness is often lateral to the sternum with a normal area between the dull space and the clavicle

Auscultation late in the course of the disease may reveal that breath sounds are enfeebled or absent If the neoplasm partially occludes a bronchus a high pitched tubular breathing is heard When a tumor closes a bronchus collapse of the part of the lung supplied by that bronchus takes place As a consequence the breath sounds are absent and both vocal and tactile fremitus are diminished or absent Like the other physical signs the rales in occurrence and nature are variable

DIAGNOSIS If possible the diagnosis is made before signs are present on physical examination The roentgenologic examination is the most important and can be diagnostically accurate in a large percentage of cases of cancer of the lung The taking of a roentgenogram after iodized oil has been injected into the bronchial tree is helpful in the diagnosis of some bronchogenic cancers

Conclusive proof of the diagnosis depends on biopsy which may be accomplished by bronchoscopy The absence of bronchoscopic demonstration of cancer of the lung however does not preclude the diagnosis of bronchogenic carcinoma for the bronchoscope cannot reach all parts of the lung In these patients examinations of the sputum for malignant cells may determine the diagnosis

A diagnosis can often be made from a biopsy of lymph nodes of the neck or axilla or other lymph nodes However when a diagnosis is thus made surgical treatment is precluded

Artificial pneumothorax either alone or combined with thoracoscopy and exploratory thoracotomy are occasionally required for a diagnosis while the lesion is still circumscribed Justification for these procedures requires an evaluation by successful and experienced specialists

Examination of sputum for cancerous tissue or cells has established the fact that the histologic type of growth can be determined frequently from cells contained in the sputum McDonald observed that the squamous type of cancer cell readily and commonly can be found by appropriate technics The oat shaped cell type is less frequently demonstrated and the ciliated columnar or goblet cell type is still less often found The failure to find cancer cells in the sputum does not disprove the diagnosis of carcinoma but is of great diagnostic aid

After the appearance of symptoms if the lesion cannot be completely excised the duration of life in most cases of cancer of the lung other than the squamous cell type varies from 5 to 9 months The squamous cell tumors usually progress more slowly

Sarcoma of the Lung Sarcomas of the lung are relatively rare A provisional classification of pulmonary sarcomas based on anatomic features has been made by Ewing *These features are as follows* (1) *diffuse spindle cell type*, (2) large round cell variety (3) peribronchial sarcoma and (4) lymphosarcoma

Sarcomas of the lung are divided clinically into the nodular and the lobar types The nodular tumors may occur as single or as multiple nodules In the lobar type a large part of the whole lobe or the whole lung is invaded Pleural effusion usually of a hemorrhagic nature often supervenes

The symptoms of primary carcinoma and sarcoma of the lung are identical

Asymmetry of the thorax and dilatation of the veins of the thoracic wall neck or arm may be present Decreased expansion of the lung involved is often relatively early in its appearance The intercostal spaces may be retracted or if effusion is present or the growth large they may bulge

Palpation confirms the irregularities in expansion Intercostal tenderness may be elicited in some instances Tactile fremitus is diminished in all surface lesions but normal in the centrally located small growths

In the small nodular growths aberrations of the percussion note may not be apparent In massive invasion of the lung there is a flat percussion note and definite resistance is encountered by the pleximeter finger In effusions of more than 500 ml of fluid the percussion note over the fluid and tumor is dull whereas just above the fluid the skodaic area of hyperresonance is sometimes found

On auscultation the characteristic findings consist of either diminished or absent breath sounds and decreased or absent vocal fremitus Rales may be present but variable accordingly if atelectasis is present Rales are constant if caused by congestion or bronchitis

The findings elicited by *physical means are never* definite enough to form the basis of a diagnosis of either sarcoma or carcinoma of the lung The roentgenologic examination constitutes the most important diagnostic procedure in cancer of the lung Whether or not the cancer of the lung is carcinoma or sarcoma is a diagnosis *to be made by the pathologist This cannot be determined* by either the palpating finger or the roentgenologic examination

Examination of the sputum for malignant cells is repeated for this is an important and specific diagnostic test Bronchoscopy is of far less diagnostic value in *sarcomas of the lung than it is in bronchogenic carcinoma* for it does not often permit of biopsy because sarcomas are usually extrabronchial

Aspiration biopsy for obtaining tissue for microscopic diagnosis may be considered after an aspiration of effusion has been done and examination of the fluid *on occasion has not revealed malignant cells Aspiration is rarely* advisable

DISEASES OF THE TERMINAL RESPIRATORY PASSAGES AND LUNGS

THE RESPIRATORY PASSAGES

The respiratory passages comprise the nose the nasopharynx and the pharynx The mouth the larynx the esophagus the two eustachian tubes and the two posterior nares open into the pharynx which thus serves as the common tract for intake of air food and drink The diseases of the air passages from the external nares up to the pharynx have been described (pp 85 to 96)

The essential anatomic structures for respiration thus begin with this common *tract* which is provided by the pharynx and extends inward through the larynx trachea bronchi bronchioles and finally the air spaces (atria) and the supporting tissues of the lungs

Respiration and Its Control *The lungs are part of a mechanism for* insuring that the tissues of the body are adequately supplied with oxygen The functions of the lungs are intimately associated with and inseparable from those of the heart and circulation All the mechanisms of respiration and circulation are integrated and presided over by the *autonomic nervous* system

A failure of proper respiration may arise therefore from defects in the supply or quality of blood to the lung membrane from defects in the supply or quality of the air or from defects in the lungs or of their chemical and nervous control

The respiratory center in the brain extends through the upper part of the medulla oblongata and probably into the pons as well Although one major and two minor centers have been described it is well to consider the central gray matter as a whole and to think of it as the respiratory center acting as a co ordinated unit It possesses an inherent rhythmicity and it controls the activity of the lower motor neurons supply ing the respiratory muscles It receives afferent impulses from the lungs via the vagus nerve and from the aortic arch and carotid sinus via the sino aortic nerves It is also influenced by impulses coming from the higher brain centers It is extremely sensitive to certain chemical variations for instance the carbon dioxide tension or the hydrogen ion content of the blood and also to the presence of various poisons such as hydrogen cyanide and carbon bisulfide

Some patients who are of a neurotic temperament and full of complaints may respond during health or during illness as a result of certain stimuli physical or emotional by overbreathing If this is sufficient to reduce the carbon dioxide con centration in the blood alkalosis ensues and in its turn may be sufficient to cause an attack of tetany The attack is due to tissue alkalosis and ends spontaneously as a result of tissue acidosis consequent on the muscular activity of the tetany Cessa tion of overbreathing assists in the return to normal conditions Alkalosis resulting from spontaneous hyperventilation may cause either tetany or effort syndrome

Defenses of the Lungs The inhaled air passes over the upper part of the respiratory tract where it is warmed and humidified The warmed air is drawn through the larynx trachea and bronchi into the depths of the lungs where the interchanges of gases can be made with the blood through the respiratory epithelium

Harmful foreign material from outside cannot easily enter much less remain in the lower respiratory passages The microbes present in the air may never reach the trachea A large number of them are trapped in the vestibules of the nose being enmeshed by the hairs which are moistened with sticky mucus and later ejected by the action of the ciliated epithelium The ciliated epithelium extends over the walls of the nasal sinuses the greater part of the nasal cavities the nasopharynx and the back of the soft palate trachea and bronchi as far down as the point where the smallest bronchioles expand into the terminal alveoli Mucus is spread in a thin even trans parent layer

The larynx protects the lower respiratory passages when need arises by its sudden reflex closure Such action is observed on exposure to irritant gases such as chlorine

Soiled mucus collecting in the interarytenoid region may excite the swallowing reflex and so help to carry off invading organisms to the stomach where most of them (but not tubercle bacilli) are killed by the acid secretion If the sensitive larynx trachea or bronchi are sufficiently irritated the cough reflex is brought into action thus foreign material is forcibly expelled a process aided by the shortening and narrowing of the bronchial tubes which is caused by the forced expiration characteristic of coughing

Organisms passing beyond the larynx are subjected to the mucociliary activity in the trachea and bronchi some may be carried away by lymphatic drainage but most will be forced back toward the pharynx

The trachea and its bronchial branches conduct air from the outside atmosphere to the air sacs of the lungs The larger bronchi divide and subdivide eventually to become bronchioles devoid of cartilage and mucous glands but with a muscular coat relatively twice as thick as that of the larger bronchi Each bronchiole ends in a lobule enclosed in a fibrous capsule with its own supply of blood vessels nerves and lym phatics The lobule forms therefore a fundamental pulmonary unit and it is the packing together of innumerable such units which with the conducting tubes makes up the lung as a whole Inside the lobule the bronchiole divides into still smaller tubes the terminal branches of which—the respiratory bronchioles—lead into an expanded space (atrium) each atrium leads into two or three air sacs (infundibula) The air sacs are studded with minute finger like protrusions the alveoli which are lined by a single layer of large flattened cells The air in these alveoli is separated from the blood capil

laries only by this single layer of cells and the tenuous walls of the capillaries The alveoli all over the lung are so packed together that any particular capillary lies between adjacent alveoli so that the blood is in close contact with air on both sides In the pulmonary meshwork between the alveoli are large numbers of elastic fibers nerve fibrils and lymphatic channels

The terminal bronchioles do not possess ciliated epithelium so that if microorganisms reach this part of the lung they will be sucked into the air sacs during inspiration and the only defense left is an inflammatory reaction in the alveolar units This is important in the pathology of pneumococcal pneumonia

CYSTIC DISEASE OF THE LUNGS

Congenital Pulmonary Cysts Congenital cysts of the lungs exist as solitary and multiple fluid or air cysts Fluid cysts have no communication with the bronchus air cysts have free communication with the bronchus

The history often reveals a susceptibility to colds and coughs Dyspnea and cyanosis may have been present from the time of birth If severe respiratory difficulties are observed at the time of birth congenital cystic disease is probable

SYMPTOMS Often the symptoms are manifested as persistent respiratory discomforts which follow an acute attack of bronchitis whooping cough or pneumonia There is cough moderate to severe with expectoration of mucoid to thick purulent sputum In the interval between these episodes of symptoms the patient enjoys a comfortable state of health if the cyst or cysts do not communicate with the bronchi If the cysts communicate with the bronchi there is a valvelike constriction at the point of communication and sudden serious or even fatal dyspnea may develop for under such circumstances the essential feature is the same as that of a bronchial fistula from any cause

Spontaneous pneumothorax may occur from rupture of the cyst into the pleural space In the presence of pneumothorax there follow the sudden paroxysmal respiratory attacks which are associated with signs of tension pneumothorax The dyspnea is critical the cyanosis severe and each convulsive respiratory struggle seems to represent the last act of the patient Thoracentesis may give temporary relief

EXAMINATION An examination by physical means may elicit the signs of local tympany or suggest a tension pneumothorax There may be a detectable displacement of mediastinal structures and descent of the diaphragm on the affected side

If a young patient has had staphylococcal pneumonia a radiolucent region in the lung is most likely an acquired cyst A cystic cavity found before any respiratory illness has occurred suggests a congenital cyst Congenital cysts usually maintain a constant size while acquired cysts or pneumatoceles tend to fluctuate in size Congenital cysts may have denser shadows (thicker walls) roentgenographically than acquired cysts Infection in congenital cysts becomes a more frequent occurrence as age increases On thoracoscopic examination the lining of a congenital cyst is smooth and glistening while the wall of a pneumatocele is black Congenital cysts of the lung persist while acquired cysts tend to disappear spontaneously

DIAGNOSIS Roentgenoscopic examination aided by means of the opaque media is usually diagnostic The diagnosis is suspected from the history and physical examination

The prognosis is good in fluid filled or air containing cysts when not complicated by infection or a one way valve communication with a bronchus

INFECTIONS OF THE LUNGS DUE TO BACTERIA

Pneumonia The conditions necessary for the inception of infection for the production of pneumonia are the implantation of virulent microorganisms or ultramicroscopic life in the terminal airways in sufficient numbers that they cannot be immediately overcome by the defensive forces of the host An obstructio o drain

age and local irritation or injury to the terminal airways hastens the inceptive processes

The reactions representing virulence of bacteria and host resistance are complex Regarding the means of passage of bacteria through the mucous membrane it is known that the absence of ciliary epithelium in the terminal airways aids the infection to become more easily established for its normal absence in the ductuli alveolares pulmonis (BNA) permits bacterial stasis in these terminal air units In influenzal pneumonia the ciliated columnar cells so situated in the larger air passages are replaced with cuboidal goblet cells resulting in the impairment of the capacity of the mucous membranes of the larger air passages to dispose of secretions effectively

In persons who have had an operation or an injury to the thorax there are often a disturbance of the cough mechanism and interference with movements of the diaphragm and of the accessory muscles of respiration and these are major mechanical factors in allowing pathogenic agents to enter the pulmonary parenchyma

In these patients the local irritation and injury are supplied by an inability to get rid of the secretions In the presence of these irritations and injuries a virulent bacterium which is present as a part of an acute respiratory infection at the time of operation and injury often can originate pneumonia

A pneumococcal infection in a particular instance depends on the individual susceptibility to the pneumococci with which the patient comes in contact provided the pneumococci possess sufficient virulence to produce disease

The pneumococcus with its type specific polysaccharide capsule when introduced into the body of the host is capable of stimulating defensive mechanisms The immune substances include agglutinins precipitins opsonins and protective antibodies Antibodies to pneumococci can be demonstrated in a large percentage of human beings It is assumed that the antibodies thus found in normal persons are the result of previous exposure to pneumococci that is of latent or manifest infection Normal carriers probably represent those who respond well to pneumococcus antigens and are therefore highly resistant to infection

The carrier state and the dosage and virulence of organisms received by the host are important factors in the production of pneumonia Equally important factors include the lowering of resistance of the individual by exposure chronic disease inanition and alcoholism Depression of protective mechanisms of the mucous membranes of the upper portion of the respiratory tract by viral and other infections is important

Pneumococcal Lobar Pneumonia The pneumococci are of the tribe Streptococceae and family Lactobacteriaceae They are gram positive frequently of lancet shape usually arranged in pairs or short chains and possess an easily demonstrable capsule

Of some 75 types of pneumococci the 32 types commonly described as causative agents of pneumonia vary considerably in distribution Types I II III V VII VIII and XIV are the most frequent in pneumonia Types I II and V usually are associated only with lobar pneumonia and are present in carriers only when the carriers have been in close contact with patients ill with lobar pneumonia In a series of 13 160 cases of pneumococcal pneumonia as recorded by Rosi the types of pneumococci in order of prevalence were I II III and VII Pneumococci associated with healthy carriers are usually type III and are present in one fourth to one half of healthy persons The consensus is that type III pneumococci are capable of causing pneumonia only in those who are debilitated or aged Type VI rarely provokes infection

PATHOLOGY The progress of the pneumonic process has been divided into three stages (1) engorgement (2) red hepatization and (3) gray hepatization The initial lesion appears to be in the alveoli of one of the lower lobes and involves the smaller air passages and alveoli Exudate fills the alveolar and bronchial spaces The infected fluid spreads in all directions by aspiration while the patient is breathing and coughing As it spreads consolidation follows until the whole lobe is solidified Some time is required before the whole lobe is involved It is thus revealed how it is possible to have engorgement red hepatization and gray hepatization present in the same pneumonic lobe of the

lung Resolution occurs as the affected tissue becomes soft and friable As the pneumonic process resolves only a small part of the exudate is expectorated the major portion is autolyzed by enzymes supplied by the degenerating leukocytes and macrophages and is resorbed

The incubation period ranges from a few hours to 1 or 2 days

SYMPTOMS The onset of lobar pneumonia is characterized by a chill fever *stitch in the side* nausea and vomiting The chill is often prolonged and is followed by a headache and generalized body aches The temperature rises rapidly to 104 to 105 F (40 to 40 5 C) Very soon there is a short unproductive cough The cough *is restrained because it enhances the pain* The pain is sharp and stabbing and its presence seems to cut the cough short In rare instances the pain is absent The face is red and the cheeks are flushed often the cheek on the affected side is more flushed *than the opposite cheek The expectoration at first is scanty it becomes blood* streaked and then brownish yellow or red in appearance The expectoration in creases as the engorgement of the lung and the cyanosis increase

The degree of cyanosis is somewhat of an index of the severity and extent of the pneumonic process Dyspnea and hyperpnea compensate only in part for the cyanosis which is due to anoxemia dependent on alveolar edema and exudate which make impossible a *free exchange of oxygen and carbon dioxide*

The pulse rate is increased usually in proportion to the increase in temperature Circulatory failure if it occurs is not primarily myocardial in origin but is more nearly related to circulatory collapse of the vascular capillary bed A sudden de crease in the blood pressure observed during the course of pneumonia is related to the tachycardia prostration and dyspnea This evidence of circulatory strain in pneumonia is likely to occur more frequently in the obese the alcoholics and the aged

A pneumonic crisis may occur within one day after onset or it may be *delayed* for two weeks or more The crisis is uncommon before the third day and rare after the twelfth A precritical rise of a degree or two of fever may occur Then within a *period of from 5 to 12 hours the temperature returns to normal (often a fall of 6 to* 8 F) After the crisis the temperature is often subnormal sweating is abundant and the patient falls asleep for several hours

There are all gradations of severity of lobar pneumonia In fulminating pneu monia there are prostration shocklike reaction and often pulmonary edema (wet pneumonia) These patients may die within a few hours from circulatory collapse In contrast there are those who have fever of brief duration with crisis within 24 hours (abortive pneumonia) Ambulatory cases occur in which the course is mild and in which the lesion is detected on roentgenologic *inspection only Central* pneu monia with location of the lesion near the hilus of the lung will often give no signs on physical examination However within a few days physical signs will appear *The involvement of one or more lobes on each side of the thorax* has been referred to as double pneumonia in which the prognosis is less favorable than with involvement of a single lobe In a massive pneumonia the bronchi of the involved region *are often* filled with exudate and the physical findings may simulate pleural effusion In migratory pneumonia the process extends through the lobes of one or both lungs In lethal instances *of the disease in which there was little or no evidence* of involve ment of neighboring lobes detectable prior to death necropsy frequently reveals such extension

EXAMINATION On *examination the face* neck and hands are hyperemic or cyanosed Dyspnea is present and there is often a herpes labialis and a flushed cheek on the side of the lesion The respirations are *increased at intervals to several times their normal rate* After consolidation has occurred unless the bronchi are filled with thick secretion or fibrinous exudate or unless pleural effusion is *present the vocal fremitus over the affected portion* of the lung is increased and a friction

fremitus also may be felt If the consolidation is deep seated the resonance may be impaired or no changes can be elicited on percussion The vocal fremitus if the larger bronchi are not plugged is increased

In the early stages the breath sounds are often enfeebled and may be suppressed When the breath sounds are present at the end of inspiration crepitant rales may be heard As engorgement advances into consolidation the breath sounds become bronchovesicular and then bronchial in type

The occurrence of persistent crepitant rales in the presence of systemic reaction cyanosis and dyspnea—any one of which alone is significant—together is almost diagnostic of pneumonia A limitation of expansion on the affected side and pleural friction rubs are often present Impairment of resonance appears or increases in intensity with consolidation If the region of consolidation is small its detection and delineation may best be facilitated by the increased intensity of tactile fremitus The demonstration of increased tactile fremitus depends on the extent of the involved space at the periphery and the experience and tactile sense of the examiner If a bronchus becomes plugged by secretions the atelectasis and absence of breath sounds with some impairment of resonance may be difficult to interpret until sudden clearing of secretions after cough or a change in position is made

Sputum and blood for cultures should be obtained before chemotherapy is started The determination of the presence or absence of invasion of the blood stream is valuable in every case Streptococci or staphylococci or other organisms may prove to be the etiologic agent and their recognition is more important for therapeutic application than the type of pneumococci The presence of such organisms as Friedlander s bacillus or *Hemophilus influenzae* may be suspected from Gram s stains The susceptibility of the invading organisms to the various chemotherapeutic drugs as well as to the antibiotic agents has made the determination of these susceptibilities a standard procedure for the proper therapeutic results

Leukocytosis is the rule in pneumonia The usual counts vary from 15 000 to 25 000 per cubic millimeter of blood Increased leukocytosis may be present with complications such as empyema or pulmonary abscess Reduction of leukocytes occurring with extension or complications or at any time during the course of the disease indicates an undesirable response and a low resistance to the disease

DIAGNOSIS Cyanosis hyperpnea dyspnea fever and chills occurring in a previously well individual and attended by physical and roentgenologic evidence of pulmonary consolidation are distinctive of lobar pneumonia In the presence of an acute exanthem meningitis or other severe systemic disease lobar pneumonia is not diagnosed until evidence of localization becomes apparent Such localization as well as the extent of the pneumonia is more precisely located by means of roentgenologic examination

The early involvement of the pleura in the lower portion of the right lung field may cause referred abdominal pain This pain may simulate the pain of cholecystitis or appendicitis so that time may be required in order to make the correct diagnosis

The organism producing the lobar pneumonia is determined by bacteriologic methods

In regard to the prognosis in pneumococcal pneumonia in recent years two groups of patients who had this disease were studied by de Beradinis and de Andino The first group of 75 patients was admitted during the period of 1937 to 1940 The second group consisted of 91 patients admitted during the period of 1946 to 1949 Comparison of the two groups showed that there was no decrease in the rate of admission of patients with pneumococcal pneumonia In the years 1946 to 1949 there was an increase in incidence of the disease in higher age groups with an increase in mean age from 39 9 years to 46 6 years There was also an increase in the occurrence of pneumococci of type IV and above with a corresponding fall in lower numbered types especially type I Physical observations of lobar con

solidation were made in 63 patients of the first group as compared with 35 in the second group The percentage of septic complications fell from 16 to 7 7 per cent probably as the result of antibiotic therapy The over all mortality rate fell from 21 3 to 6 59 per cent The majority of the patients who died had the complicating features of chronic degenerative disease which influenced the mortality rate un favorably in patients aged more than 40 years

COMPLICATIONS Empyema and pulmonary abscess are the commonest complications Empyema is suspected if there are (1) a lysis instead of a crisis of the fever (2) recrudescence of fever (3) a continuance of the increased leukocytosis, (4) sweats and a progressive inanition and (5) the physical or roentgenologic evidence of free or encapsulated pleural effusion

Delayed resolution bronchiectasis pulmonary fibrosis pericarditis meningitis, endocarditis hepatitis parotitis and purulent arthritis are of rare occurrence when sulfonamide or antibiotic therapy is employed

Bronchopneumonia Bronchopneumonia is a diffuse process not often caused by the pneumococci It is not lobar in distribution A bronchial pneumonia differs from the so called atypical pneumonias by the fact that etiology can be determined The disease most frequently is caused by beta hemolytic streptococci Other causative organisms are *Hemophilus influenzae Staphylococcus aureus* gram negative micrococci and *Streptococcus viridans* Bronchopneumonia in adults is usually secondary to an acute infection of the upper part of the respiratory tract influenza measles or chronic disease It tends to be most frequent in early and late life

SYMPTOMS Since bronchopneumonia is almost always secondary to a preceding illness the symptoms are merged with those of that illness The initial symptoms of bronchopneumonia are fever cough and dyspnea Rusty sputum is less frequent than in lobar pneumonia There may be a chill or a chilly sensation Pain develops immediately The fever is of the irregular daily fluctuating type Prostration and signs of anoxemia vary considerably and while they tend to be less in bronchopneumonia than in lobar pneumonia nevertheless a marked cyanosis may develop which is indicative of a severe process The symptoms of the disease may fail to have a respiratory localization and thus there may be no respiratory symptoms Whether or not there are respiratory localization and symptoms the fever tends to terminate by lysis The duration of the disease is exceedingly variable and all symptoms may merge into those of a complication Complications are less frequent in bronchopneumonia however than in lobar pneumonia

The presence of acute respiratory symptoms and fever in a patient who has had influenza measles or any respiratory infection which has reached epidemic proportions is accepted as pneumonia bronchial in origin until proved not to be of such origin

EXAMINATION The physical examination reveals findings varying from those present in acute bronchitis to those of lobar pneumonia At times bronchopneumonia may involve almost an entire lobe of a lung The presence of several localized regions of crepitant rales even if the large part of one lobe is involved is characteristic of bronchopneumonia The percussion findings depend on the size of the pneumonic patches in the lungs Often there are no definite changes to percussion or breath sounds

DIAGNOSIS A history of previous respiratory infection physical signs and roentgenologic evidence in the thorax are conclusive

When bronchopneumonia occurs secondary to chronic disease particularly in the advanced decades of life high mortality rates are expected Following epidemics of measles respiratory infection and influenza a high mortality rate is the rule Increasing age, marked extent of pulmonary involvement bacteremia alcoholism inanition and debilitating disease are all factors making for grave prognosis

Postoperative Pneumonia This disease is bronchopneumonia occurring soon after a surgical operation The predisposing causes of postoperative pneumonia (bronchopneumonia) are listed by Whipple as follows (1) inflammations in the upper part of the respiratory tract present at the time of surgical operation (2) vasomotor changes causing a congestion of the pulmonary vessels (3) factors during and after the operation which inhibit the normal thoracic and abdominal respiratory movements and favor atelectasis and hypostasis in the lung (4) local or general infections elsewhere than in the respiratory tract (5) a lowered natural or acquired immunity to organisms which can incite pneumonia and (6) increase in the virulence of the inciting organisms for instance the presence of a virulent strain of streptococci causing endemic sore throats

Postoperative pneumonia often begins as atelectasis with pneumonia developing in the collapsed portion of lung The symptoms are those of bronchopneumonia The onset of symptoms may be as early as the second postoperative day or delayed until the patient is out of bed The physical findings are likewise those of bronchopneumonia

The bedside roentgenologic examination is used as guesswork for diagnosis until the patient is able to be moved for physical examination and the regular roentgenologic examination

Post traumatic Pneumonia Thoracic injuries especially fractured ribs which cause interference with respiration are frequently associated with pneumonia Interference with respiration appears to be the most important single factor in the occurrence and localization of pneumonia or atelectasis in post traumatic as well as postoperative patients Interference with respiration also accounts for an occasional pneumonia after severe head injuries

The etiologic agent in post traumatic pneumonia is often a pneumococcus A rib fracture in an elderly person followed by infection with pneumococcus type III may have a serious prognosis Streptococci staphylococci or other organisms are often the causative microorganisms and they too cause a serious pneumonia in a patient of any age

The symptoms are often obscured by those of the original injury The expectoration of blood often indicates a pulmonary hemorrhage A fever and leukocytosis associated with respiratory distress and hemoptysis are significant manifestations

Following trauma intrapulmonary hemorrhage may be the cause of physical signs like those associated with bronchopneumonia The physical signs after trauma are usually scattered with only at times those of consolidation of a lobe

A presumptive diagnosis can be made if there are no evidences of intrapulmonary hemorrhage on roentgenologic examination

Prognosis is variable depending on the resistance of the host in relation to age chronic disease alcoholism and the virulence of the invading organism

Pneumonia Due to Unusual Bacterial Agents Infrequent agents causing pneumonia include bacilli such as *Hemophilus influenzae Hemophilus pertussis Bacillus pyocyaneus* (*Pseudomonas aeruginosa*) *Bacillus anthracis* and *Pasteurella pestis* cocci such as *Gaffkya tetragena* meningococci and *Neisseria catarrhalis* Pneumonia may also be associated with brucellosis typhoid fever diphtheria and rickettsial diseases

Acute Pneumonitis The term acute pneumonitis is used by Adamson (in Myers and McKinlay) to indicate acute inflammation in the lung that is not only endobronchial but involves the parenchyma as well The parenchymal involvement in acute pneumonitis is not sufficient to produce signs or symptoms of pneumonia The term thus includes many separate etiologic entities yet if the definition of the term as it is given by these authors is adhered to the underlying pathologic condition is definite and precise

Adamson has arranged a convenient classification of pneumonitis which is essentially as follows (1) common cold pneumonitis (2) influenzal pneumonitis (3) obstructive pneumonitis (4) exacerbation pneumonitis

1 **Common Cold Pneumonitis** Gross pulmonary infiltration is found on roentgenologic examination of the thorax in many patients who have insignificant symptoms Often there is a history of a *common cold or slight infection in the upper part of the respiratory tract* A *follow up on these patients shows complete resolution in the course of a few weeks*

Physical signs are usually absent

2 **Influenzal Pneumonitis** The pneumonitis following influenza is essentially inflammatory and that following infection of the *upper part of the respiratory tract is commonly obstructive In most instances of influenza the respiratory symptoms* commence *after* the first day after the onset whereas in infections of the upper part of the respiratory tract the inception is with respiratory symptoms

Influenza is characterized by the sudden onset of headache muscle pains anorexia chilliness weakness depression and general malaise If there is a localization of the discomfort it is the backache and aching legs *There may be signs or symptoms of respiratory inflammation varying from pharyngitis to pneumonitis* or in some a definitely manifested pneumonia

There is a widespread bronchitis accompanied by retrosternal pain and a burning dry harassing cough with scanty sputum There may be coarse rhonchi diffusely spread throughout both lungs Crepitations after coughing are commonly found about the bases which *indicate definite parenchymal infiltration*

On roentgenologic examination of the thorax there is a peribronchial infiltration which spreads out into the parenchyma from one or both roots The commonest site is in the medial half of the base the cardiophrenic angle being frequently involved The extension may be into the middle third and not infrequently into the upper lobe In most cases the involvement is unilateral but both lower lobes may be involved at the same time or in sequence Migration from place to place *is common*

Characteristically influenza does not produce leukocytosis and *if the total* leukocyte count is more than 10 000 per cubic millimeter the pulmonary *infection* is due to or complicated by some pyogenic organism The common occurrence of a relative bradycardia in influenza is of value in the recognition of influenzal pneumonitis

3 **Obstructive Pneumonitis (Infection Atelectasis)** An obstructive *pneumonitis may arise when there is an abnormal respiratory* secretion It is *characterized* by sudden onset of pleural pain *in the axillary line associated* with shortness of breath Constitutional signs are mild or absent *The physical and* roentgenologic signs are those of pulmonary collapse elevation of the *diaphragm* or displacement of the mediastinum

This condition requires treatment primarily directed to relief of obstruction

4 **Exacerbation Pneumonitis** Pulmonary tissue damaged by previous inflammation may have as a consequence an impairment of natural drainage Reduced natural drainage is *conducive to accumulation* and *stagnation* of secretion and retention of *infected material* As the result of poor *drainage* local inflammation occurs which is a pneumonitis A *variety of clinical effects may be produced* depending on the relative participation of the two *factors obstruction* and inflammation and the extent of the process

There is a history of colds cough and purulent expectoration In a *few cases* there is the history of attacks with pain and fever In the intervals between *attacks* the patients are free from symptoms or chronic cough may persist The *attacks* are always localized and repeated in the same side

The physical signs and roentgenologic findings during any such attack are similar in a general way to those of any other form of pneumonitis

There may be signs of chronic infection enough to suggest chronicity Such signs are clubbing of fingers thoracic asymmetry reduced movements dullness or coarse rales which do not clear after coughing

Roentgenologic observations may suggest chronic rather than acute changes

Pulmonary Abscesses In a patient in whom the presence of a pulmonary abscess is suspected the history of a surgical operation particularly on the mouth nose or throat or of a recent attack of pneumonia is significant Pulmonary abscesses may follow surgical procedures which are prone to give pulmonary infarcts such as operations on the pelvis and hernial repairs They may occur during the course of a septicemia such as endocarditis or furunculosis History of an attack of choking while swallowing food or an attack of cough following aspiration of some other object is significant from the standpoint of abscess due to foreign body In some patients who have pulmonary abscesses there is no history of pre existing conditions such as those just mentioned This is particularly true of pulmonary abscesses in children following aspiration into an air passage of a foreign body which supposedly was coughed up

The pyogenic abscess is usually caused by the pneumococcus *Staphylococcus aureus* and the hemolytic streptococcus Anaerobic organisms which normally inhabit the upper air passages of adults and are especially numerous in the presence of carious teeth gingivitis pyorrhea and diseased tonsils cause putrid abscesses Saprophytic streptococci spirochetes fusiform bacilli and gram negative bacilli in combination acting in symbiosis can produce lung abscesses

A pulmonary abscess may arise from an infected embolus causing infarction and secondarily abscess formation In other instances lodging of a sterile embolus produces infarction organisms are aspirated from the mouth nose and throat and infect the infarcted region in the lung and thus an abscess is formed In some instances both processes are present namely an infected embolus and the aspiration of infected material from the nose throat and trachea

The prime factor which favors the chances of abscess formation after operations on the nose mouth or throat is the use of general anesthesia for the operation The general anesthesia abolishes the cough reflex and thus permits aspiration of infected material from the upper air passages

Pulmonary abscesses often result from the aspiration of foreign bodies into a bronchus Abscesses due to aspirated foreign bodies are more frequently found in the lower lobes especially the right Once the foreign body is lodged in the bronchus it ulcerates through the wall of the bronchus into the pulmonary tissue and abscess formation ensues The object for abscess formation may be of intrapulmonary origin for instance Vinson called attention to calcifications produced by the first infection type of tuberculosis in hilar lymph nodes which ulcerated into bronchi and were aspirated into their ramifications causing abscess Other causes of pulmonary abscess are obstruction of bronchi by tumors and pneumonia

SYMPTOMS A chill followed by fever similar to that observed in pneumonia may be the first manifestation of pulmonary abscess In the beginning of the disease cough and expectoration are slight or absent As the condition progresses the cough begins and increases in severity Pleural pain and the sudden expectoration of a quantity of sputum with a foul odor are common manifestations Abscesses which follow operations and aspirations from the mouth and throat usually are of sudden and severe onset There are profuse sweating and prostration Pulmonary abscess does occur with less severe initial symptoms being manifested only by a general aching of the body and malaise Spitting of blood varying in amount from streaks in the sputum to fatal hemorrhage occasionally occurs even in those who have mild symptoms The symptoms of multiple abscesses due to septic emboli often are masked by the symptoms of the main disease During the course of septicemia or pyemia the first symptoms of a pulmonary abscess or abscesses may be

thoracic pain cough and expectoration No *one symptom or combination of* symptoms is pathognomonic of pulmonary abscess *since all may be caused by* other diseases

EXAMINATION Inspection often reveals clubbing of the fingers and toes sometimes within a few weeks after onset of the abscess Hypertrophic pulmonary osteoarthropathy develops in some instances Limited movement of the thoracic wall on respiration often occurs on the affected side and the phrenic wave sign is decreased or absent If the abscess has an acute severe onset the patient appears seriously ill

If the abscess is small no abnormal signs are elicited by palpation If the abscess is large with extensive inflammation of the adjacent pulmonary tissue tactile fremitus is increased except in the presence of pleural exudates when it is diminished or absent The reflex protective mechanism consists of a rigidity of thoracic muscles which can be elicited and palpated by tapping on the chest over the affected area with the end of the stiffened fingers

The percussion note is normal if the abscess is small but when it is large and near the surface a dull to a flat sound is elicited Over large cavities near the surface of the lung a tympanitic sound may be heard when the cavity or cavities are empty

On auscultation the breath sounds are normal if the region of disease is small but as it becomes larger bronchial breathing appears If large cavities are near the surface and are partially or totally empty amphoric breath sounds may be heard Changes in the spoken and whispered voice sounds *depend* on the extent and situation of the disease Voice sounds are normal when normal lung tissue is superimposed over the abscess but are heard distinctly over the areas where bronchial breathing is elicited and if large empty cavities exist pectoriloquy may be present Rales are usually heard when the region of disease is large and near the surface These may be consonating if cavities are only partially filled

DIAGNOSIS The foregoing symptoms and physical signs may be definite enough for the purpose of diagnosis when interpreted and correlated with the history

Roentgenologic examinations often determine the location and extent of gross disease although some abscesses are not located by this means Large abscess cavities when completely filled with pus present no fluid level their outlines may not be seen so that their presence is not detected on roentgenologic examination If evacuation of the abscess cavity or cavities as by *postural drainage can be accomplished then iodized oil can enter after which the roentgenogram affords good evidence as to size and location of cavities Endoscopic inspection* of bronchi *should always be made if there is doubt as to* either the presence or the etiology of an abscess

Pulmonary abscesses may occur in tularemia brucellosis and amebiasis Inasmuch as tuberculosis may coexist a search is made for tubercle bacilli and if there is doubt guinea pig inoculation is performed

A single abscess may heal spontaneously in 1 of every 4 cases In the remaining 3 cases the prognosis is guarded The poor prognosis in many is due to conservatism and the lack of surgical skill Spontaneous recovery rarely occurs *if the disease* is allowed to persist more than a few weeks *Surgical treatment as soon as the* diagnosis is made *markedly improves the prognosis*

Pertussis (Whooping Cough) The genus *Hemophilus pertussis* (whooping cough *bacilli*) *type Hemophilus influenzae* belongs to the family of Parvobacteriaceae

Strains *of Hemophilus pertussis* on primary isolation constitute a single antigenic type They are gram negative The organisms can be identified serologically *by the* agglutination test

The distribution of pertussis is *cosmopolitan* The disease *is endemi* ... ies

occur at intervals of two to four years in the more thickly populated areas beginning in winter or spring and lasting throughout the summer months In the South the disease spreads during April and May in the North January and February are the peak months Pertussis is distinctly a disease of early life except in some women particularly in the older age groups Infants less than 6 months of age are especially susceptible and among them the disease has a higher mortality The communicability rate is high particularly in family exposures in which it is from 75 to 90 per cent

IMMUNITY Although an attack usually confers lasting immunity second attacks do occur

Active immunity against the disease can be induced by the injection of an antigenic strain of the organisms Passive protection may be given to an infant by the injection of convalescent serum hyperimmune human serum or immune rabbit serum It is not generally transferred transplacentally but can be produced in this manner by actively immunizing the pregnant mother during the third trimester of pregnancy

A profuse catarrhal inflammation of the epithelium in the larger air passages is always present Purulent exudate and an excess of mucus accumulate in the lumina of the smaller bronchi and bronchioles Peribronchiolitis is common extending from the hilus outward along the blood vessels which accompany the bronchi to the middle or outer portions of the lungs

The tracheobronchial lymph nodes are usually enlarged during the active and early convalescent stages of the disease and produce the characteristic hilar lesion commonly observed on roentgenologic examination

In addition to the pulmonary lesion produced by *Hemophilus pertussis* secondary invading organisms may cause purulent bronchitis and even lobar pneumonia Bronchiectasis may develop early in the course of the disease but it is commoner as a sequela Emphysema is almost always present blebs on the surface of the lung being observed in practically all cases in which death ensues Changes in the brain are described as circulatory degenerative and inflammatory Meningitis and encephalitis result from inflammation

SYMPTOMS The incubation period is from 10 to 14 days The clinical course consists of three stages the catarrhal paroxysmal and convalescent each lasting approximately 2 weeks Spasmodic coughing may occur for several weeks or months longer

The first symptom commencing with the catarrhal stage is a mild nocturnal cough which soon becomes diurnal The cough develops as that of a bad cold but the appetite fails as the cough increases in severity and there is a suffusion of the conjunctivae with edema of the lower eyelids

Toward the end of the second week the paroxysmal stage commences The cough becomes a series of explosive efforts of violent expiratory coughs during which inspiration is impossible Finally inhalation is performed through a markedly spastic larynx and there is a rather loud high pitched inspiratory noise called the whoop During the period preparatory to the whoop and soon thereafter the veins of the neck and forehead are prominent the face becomes congested and cyanotic and mental confusion results In infants the characteristic whoop may not occur but choking cyanotic attacks are common

The convalescent stage commences after a month of coughing and the paroxysms diminish in frequency and severity The appetite improves If a cold develops during convalescence all the symptoms may reappear and continue for several weeks more if a superimposed bronchitis remains active

Pertussis may be extremely mild and may last only a few days without definite paroxysms whoop or vomiting

At any time during the course of the disease complications may supervene and the symptoms and physical findings are accordingly changed

Bronchopneumonia is a serious complication Atelectasis bronchiectasis pulmonary fibrosis and emphysema either vesicular or interstitial develop in many instances when the disease is severe Pneumothorax and empyema may occur

Subcutaneous emphysema occurs when air from ruptured surface blebs escapes by way of the mediastinum into the tissues of the neck and thorax In whooping cough in contrast to spontaneous pneumothorax subcutaneous edema is a serious complication and implies a fatal outcome

Convulsions are most commonly limited to infants Encephalitis epilepsy mental retardation visual defects temporary paresis and hemorrhages of the brain are described

Secondary mechanical effects attributed to the strain of coughing are conjunctival hemorrhages epistaxis ulcer of the lingual frenum intussusception hernia and rectal prolapse Often as the result of coughing in adolescents there is soreness of the muscles of the lower part of the thorax and particularly of the upper part of the abdomen

EXAMINATION The findings on examination by physical means depend on the stage of the disease and the presence or absence of complications when the patient is examined During the course of mild to moderate degrees of severity of the disease there may be very few or no significant physical findings In the severe instances of the infection there are various sorts of rales throughout both lungs The presence of crepitant rales in any part of the lung often signifies the onset of a bronchial pneumonia in the region where the rales are present

A progressive increase in the total leukocyte count occurs during the late catarrhal or early paroxysmal stage These cells may increase to 15 000 to 45 000 per cubic millimeter of blood as the result of lymphocytosis

DIAGNOSIS A history of definite exposure is of much diagnostic value A modified type of the disease may be expected when vaccination fails to protect completely

Hemophilus pertussis may be isolated by cultures from the respiratory tract During the catarrhal period positive cultures may be expected in from 70 to 90 per cent of the cases

The diseases which may produce symptoms simulating those of pertussis are infection of the adenoids or nasal sinuses allergic reactions and infection of the trachea with *Hemophilus influenzae* Parapertussis resembles mild pertussis and can be differentiated only by culture

The mortality rate is about 25 per cent during the first year of life and 10 per cent during the second year The disease is seldom fatal after the fifth year

Hemophilus Parapertussis (Bacillus Parapertussis) *Hemophilus parapertussis* is a short ovoid gram negative nonmotile bacillus which in many respects resembles *Hemophilus pertussis* It is the cause of parapertussis, a disease which resembles mild pertussis from which it can be distinguished only by bacteriologic methods

The incubation period of the disease in man is from 6 to 15 days The onset is similar to that of whooping cough but may be more abrupt The cough is less severe than that of whooping cough but is spasmodic and is sometimes followed by a whoop Vomiting is less frequent than in whooping cough The entire course of illness is from 1 to 3 weeks The disease often resembles tracheitis

It is not known whether one attack of parapertussis confers lasting protection An attack of parapertussis does not confer immunity against pertussis nor does an attack of pertussis confer protection against parapertussis

Pulmonary Tuberculosis Primary Phase In the majority of those who have pulmonary tuberculosis there is a known exposure to the disease In children often the exposure has been prolonged Prolonged exposure to contagious tuberculosis is not necessary for the acquisition of a sufficient number of tubercle bacilli to become infected for an exposure of seconds of time under proper conditions may be adequate for the transfer of large numbers of

Tuberculous infections are divided into the primary phase or the first infection and the secondary or reinfection tuberculosis

The National Tuberculosis Association defines the primary phase of a tuberculous infection as the pathologic processes which follow directly and uninterruptedly the first implantation of tubercle bacilli

In the first infection by tubercle bacilli the organisms gain entrance through the nose mouth, eyes abrasion of the skin or through the genital tract The bacilli are phagocytosed by neutrophilic cells After the bacilli are in these cells they may be inactivated or deposited in the tissues and are thus focalized These neutrophilic cells containing tubercle bacilli may enter the blood and may be carried about in the blood stream to lodge in those parts of the body most richly supplied with fine capillaries Thus lesions may be originated as focal disease in the lungs bones joints lymph nodes and meninges

The first infection of tuberculosis usually is caused by inhalation of tubercle bacilli The primary infection focus consists of a small tuberculous pneumonia often situated in the lower parts of the upper lobes or in the upper parts of the lower lobes near the pleura The center of the focus undergoes caseation rapidly (parenchymal focus) As caseation proceeds tubercle bacilli are liberated and carried through the lymph channels to the regional bronchopulmonary lymph nodes

The primary complex heals promptly However in children exposed to prolonged infection it may not heal A failure to heal may result in a parenchymal lesion which may ulcerate into a bronchus and permit of bacillary dissemination through the bronchial system The primary hilar lymph node foci may ulcerate into an adjacent bronchus and cause a fatal tuberculous bronchopneumonia

SYMPTOMS In the first infection type of tuberculosis the symptoms simulate a bad cold or influenza The febrile reactions of this type of tuberculosis are of short duration usually lasting not longer than 10 days or 2 weeks The temperature ranges between 99 and 101 F (37 2 and 38 3 C)

It is only when the primary infection fails to heal and forms a parenchymal pneumonia which ulcerates into a bronchus or when the hilar lymph nodes ulcerate into a bronchus that the primary infection causes a serious disease such as a bronchopneumonia These primary bronchopneumonias may cause death The serious manifestations of a primary infection may occur at any age and are not limited to children

EXAMINATION In the usual instance of first infection tuberculosis the physical examination is of no value in the detection of the tuberculous process in the lungs of children or adults

It is usually stated that when healing of the primary complex occurs two signs are left The first sign comprises the lymph node foci which partially or entirely heal and become calcified Even though calcification has occurred in the lymph nodes viable tubercle bacilli may remain The second and the most definite sign left to indicate the presence of the primary complex is the fact that the body has become allergic to tubercle bacilli or their metabolic products which can be demonstrated by positive tuberculin reactions

Positive findings on roentgenologic examination are present in but a few of those who are affected with a first infection type of tuberculosis

Several years after the first infection type of tuberculosis there may appear on the roentgenograms sharply outlined shadows in the pulmonary parenchyma or the hilar region which represent dense deposits of fibrous tissue calcium or bone as the result of first infection tuberculosis This is a Ghon lesion or complex

Tuberculosis as the cause of calcification in the lungs and hilar region is responsible in only one half the instances in which calcifications are visualized by roentgenologic examinations (see Calcifications of Lungs page 617)

Reinfection Phase Following the advent of the primary infection with pulmonary tuberculosis a latent period intervenes This latent period may last indefinitely if reinfection does not occur The latent period is the time supervening between the primary infection and the reinfection and the development of symptoms from the reinfection

The sources from which the reinfection is derived are always listed as exogenous and endogenous reinfections The exogenous source is from anywhere that live virulent tubercle bacilli may be obtained

There are four sources of *endogenous* reinfection (1) a primary pulmonary focus of the lung may break down and discharge bacilli into other parts of the lungs (2) a caseated primary regional lymph node focus may rupture into a bronchus causing a tuberculous aspiration bronchopneumonia (3) an active primary regional lymph node focus situated inside or outside of the lung may send tubercle bacilli to the lung or other parts of the body by way of the lymphatics and the blood stream (4) small apical foci which have developed as a result of early hematogenous dissemination frequently become progressive ulcerate and discharge bacilli into the bronchi

Three forms of tuberculosis are discussed here

1 **Miliary Tuberculosis** Miliary tuberculosis is caused by the discharge of the contents of a tuberculous lesion into either the blood or the lymph streams Hematogenous dissemination may occur during the active phase of the primary infection type of tuberculosis or as a terminal miliary spread in the reinfection type of the disease It is commoner in those who have not had symptoms of tuberculosis than in those who have had symptoms

Many of the tubercle bacilli that reach the blood stream are filtered out by the lungs kidneys peritoneum bones and the central nervous system However in 4 cases of each 5 the main localization is in the lungs due to the extent of the capillary bed here

The symptoms commence with weakness malaise and fever which increase in severity The temperature ranges from 101 to 104 F (38 3 to 40 C) The respiratory rate is normal at first but increases and cyanosis of a marked degree develops in the late stage of the disease Cough is slight in the beginning but becomes distressing as the fever and cyanosis develop *The sputum is scant*

On examination the patient appears seriously ill Results of examination of *the thorax by physical means are negative* Roentgenologic examination of the thorax usually is not helpful in the beginning of the disease Ophthalmoscopic examination may reveal the presence of tubercles in the retina

In the presence of a more continuous fever a greater failure of health a more rapid loss of weight and a more severe weakness in a patient known to have tuberculosis a presumptive diagnosis of miliary tuberculosis can be made In all others signs of localization usually in lungs must be awaited Tubercles may be found in the retinae and may be the first definite diagnostic sign *Often in severe* fulminating instances of the disease the diagnosis is not made during life except in those known to have tuberculosis In the late stage in approximately one half of those who have miliary tuberculosis a large number of small shadows appear in the *roentgenograms* of the lungs and may be the first definite diagnostic finding

2 **Tuberculous Pneumonia (Galloping Consumption)** A tuberculous pneumonia may result from a lesion of the primary phase eroding into an air passage and discharging its contents into the lumen A discharge of the tubercle bacilli into the bronchi may originate from a chronic reinfection type *of lesion* and cause a pneumonia Occasionally a small focus of either the primary phase or the reinfection type of the disease may rupture into a blood vessel so that the bacilli

are disseminated into that part of the lung which this vessel supplies This may involve a part of a lobe, a whole lobe or an entire lung When large regions of pulmonary tissue are affected the hypersensitiveness caused by the tuberculo protein aids in a necrosis of the tissues and a rapid development of pulmonary cavities

SYMPTOMS The disease frequently occurs in those who seemingly have been in good health At the onset the symptoms are those of lobar pneumonia or of bronchopneumonia In some instances the onset is sudden commencing with chills high fever and pain in the thorax

The onset may be more slowly progressive with cough fever aching of the body anorexia and malaise These symptoms soon begin to be more severe There is a rapid loss of body weight and the cough is severe Sputum appears increases in amount is yellowish or greenish and streaked with blood often profuse hemorrhages occur

EXAMINATION The patient is seriously ill The cheeks are flushed and sunken and the eyeballs are soft and retracted The respirations are increased profuse sweating and chills are regularly observed

On examination of the lungs palpation percussion and auscultation reveal the findings of pneumonia In many instances the signs of cavitation appear at short intervals after the disease has begun Reports on examination of sputum are positive for large numbers of acid fast bacilli which on culture and guinea pig inoculation prove to be tubercle bacilli

DIAGNOSIS The diagnosis is made by recovery of tubercle bacilli from the sputum The physical and the roentgenologic examinations reveal pneumonia

In most cases of massive tuberculous pneumonia the prognosis is grave as death usually ensues within a few weeks There is no specific treatment

3 ***Chronic Forms of Pulmonary Tuberculosis*** In the chronic forms of tuberculosis the organisms may be either the human or the bovine type (rarely avian according to Feldman) Chronic ulcerative and progressive pulmonary tuber culosis is a reinfection type of the disease in adults and occasionally in children

There is no good evidence that race and nationality predispose to the develop ment of pulmonary tuberculosis The incidence of the disease in the Negro and the American Indian probably is due to poor sanitation which predisposes to con tagion rather than reduced resistance The members of both races respond to tuberculosis control measures

The development of pulmonary tuberculosis following whooping cough measles pneumonia or pregnancy is probably coincidental to an environment full of the germs of tuberculosis The development of pulmonary or any form of tuberculosis in the diabetic patient is serious

Trauma does not cause tuberculosis The discovery of pulmonary tuberculosis in a recently injured individual is an accidental finding and has nothing to do with the injury

Chronic pulmonary tuberculosis frequently results from hematogenous spread of tubercle bacilli Insufficient numbers are released to cause a general miliary tuberculosis consequently fewer bacilli are deposited in various places in the body and the body is more able to cope with them The initial lesion the tubercle is a small lesion These smaller lesions extend into the air passages particularly the smaller bronchial ramifications after which sputum containing tubercle bacilli is expectorated shadows on the roentgenograms appear and other symptoms become evident

Chronic pulmonary tuberculosis may be produced by exogenous reinfections by inhalation of tubercle bacilli or by ingestion of the bacilli into the digestive tract

from which they are taken up by the lacteals They reach the blood stream and lodge in the lungs

From either endogenous or exogenous reinfections bacilli may reach the larynx and pharynx by way of the trachea may reach the digestive system through the esophagus and may reach the urogenital organs the nervous system and bones through the blood stream

Chronic pulmonary tuberculosis is most commonly situated in the upper lobes of the lungs but may appear anywhere in the lungs There is a strong tendency for such lesions to undergo remissions and exacerbations and at any time they may be permanently arrested or may continue to progress and cause damage beyond repair

SYMPTOMS A history of exposure to those who have open tuberculous lesions capable of discharging bacilli is obtained from less than one half of the patients who have active pulmonary tuberculosis

The consensus is that the allergic reaction to the tuberculoprotein is responsible for the acute manifestations of tuberculosis However the greater part of the symptoms of pulmonary tuberculosis are due to a toxemia The toxemia is manifested by its effects on the sympathetic nervous system and endocrines particularly adrenals and thyroid Pottenger enumerated the symptoms due to toxemias thus (1) malaise (2) lack of endurance (3) loss of strength (4) nervous instability (5) diminished digestive activity (6) increased metabolic rate (7) loss of weight (8) increased pulse rate (9) fever and (10) night sweats Any one or any combination of these symptoms may be present in tuberculosis Every one of these symptoms may be present in a patient who does not have tuberculosis A patient may have advanced active tuberculosis and not have any of these symptoms

Malaise is so common a complaint in those who are well or sick that it has little or no diagnostic significance at any time If present it is due to toxemia

A *lack of endurance* is not significant if it is detached from the past performance of the patient It is significant if the patient is unable to endure more than a small part of the day s work which has been performed in the past without exhaustion

The *loss of strength* is a comparative estimation in regard to past performance without fatigue Patients may state that they are unable to do work requiring the same strength that they did easily in former years In the young man or woman this may be significant In aging men and women who do not have pulmonary tuberculosis this is their most common complaint

Diminished digestive activity is often an early symptom in pulmonary tuberculosis It is accompanied by a loss of body weight Pulmonary tuberculosis is one of the common causes of loss of weight A loss of several pounds during a few weeks is always significant Indigestion often appears intermittently Digestive disturbances may be severe in one and mild or even absent in another who has tuberculosis

The *pulse rate* in pulmonary tuberculosis as in normal health, varies considerably Fever is attended by an increased pulse rate

Fever is a significant symptom of early pulmonary tuberculosis The presence of fever is determined by recording the temperature every 2 hours for a period of 4 or 5 days Elevations of temperature do not always occur at the same time of day or night on successive days The common opinion that the fever appears only during the afternoon is often correct If the temperature is taken by mouth the thermometer should be held under the tongue for 10 and preferably 15 or 20 minutes To be of significance it must be 99 F (37 2 C) or more for men and 99 6 F (37 5 C) or more for women and should not show wide variations from hour to hour In those who have a wide range of fluctuation the morning temperature may be as low as 97 F (36 1 C)

Temperature greater than 99 F may occur intermittently for instance elevated for a few days then absent for a time Three or 4 weeks of observation may be necessary in some instances to be certain fever is present There is usually a slight elevation of temperature associated with the menstrual period or at the time of ovulation a week or two before the period Slight elevations in temperature may occur after exercise

Night sweats are of frequent occurrence These sweats occur after midnight while the patient is sleeping soundly

Tuberculous patients may complain of *huskiness* or *hoarseness* of the voice without detectable changes in the larynx Huskiness of the voice occurs as a part of general weakness and may accompany any low grade fevers from any cause

Tuberculosis is one of the causes of *cough* Acute and chronic respiratory infections air contaminants allergy mitral stenosis and intrathoracic pressure are examples of some of the common causes of coughs Many tuberculous patients who have a cough attribute all the symptoms to it They feel no pain or other sensation within the thorax but have an almost constant *tickling in the throat* This symptom is a reflex one In early tuberculosis the cough may be nonproductive

Thoracic pain is commonly present in those who have tuberculosis In the absence of pleural pain which is manifested in association with inspiration a person who has tuberculosis frequently complains of pains about the shoulders and in the thorax Pains of this distribution however are often complained of by those who do not have tuberculosis

The spitting of *blood streaked sputum* may be the subject of complaint in tuberculosis as well as in many other conditions of the nose and throat Hemorrhages varying in amount from 1 dram to the point of exsanguination and death may occur Death results from pulmonary hemorrhage in one third of tuberculous patients Contrary to former-day teaching that blood streaked sputum often indicated pulmonary tuberculosis it is now known that hemorrhage from the lungs occurs much more frequently in nontuberculous conditions such as bronchiectasis than it does in pulmonary tuberculosis Patients are not fearful of small hemorrhages for there are those who give histories of hemorrhage for months or a year or more before reporting for examination

A hemorrhage may be the first manifestation of a tuberculous lesion which is so small that physical signs are not present Many patients however pass through all the stages of tuberculosis until death without hemoptysis The presence of hemorrhage requires the discovery of its cause through examination of nose throat lungs and often the esophagus and the cardia of the stomach

Sputum if present in the early stages of tuberculosis often is in small amounts thin and clear and resembling saliva As the disease advances the sputum increases in amount becomes thicker and attains a yellow or green coloration

Frequent bad *colds* and *cough* which persist for weeks are manifestations of a failure in health There are many causes for such bad colds but among these causes pulmonary tuberculosis is an important one

A preceding or a present attack of pleurisy is often the first clinical manifestation of tuberculosis

Examination On examination the patient often appears to be in good health or there may be evidence of loss of body weight and flabbiness of the muscles As the disease progresses an extreme state of emaciation may develop Often there is a detectable decrease in the expansion of the thoracic wall over the region of disease The excursion of the diaphragm is limited hence the phrenic wave (Litten s sign) is limited in range of excursion

Often a patient who has pulmonary tuberculosis has a flushed condition of the cheeks The flushing may appear only at certain times of the day and is not synchronously present with fever The reddened cheeks may be unilateral or bilateral

corresponding to the side of the lesion or lesions The patient may complain of a hot sensation of the ear or the side of the face without the appearance of flushing The burning and the flushing are due to dilatation of the blood vessels to the regions produced reflexly by the stimuli passing upward over the vagus and peripherad over the fifth cranial nerve

When the disease attains the moderately or far advanced stage tactile fremitus is present The percussion note is dull or flat over the involved portions Auscultation reveals changes in breath sounds varying from bronchovesicular to bronchial breathing The whispered and spoken voice sounds are more audible over the tuberculous lesions than over the normal lung Bronchophony may be elicited over sites of solidification and pectoriloquy over the empty or partially filled cavities Rales of the fine or moderately coarse variety are heard over the regions of disease which have reached the moderately or far advanced stage Rales of a metallic or resonating quality are heard over cavities

DIAGNOSIS OF PULMONARY TUBERCULOSIS The following discussion is essentially an adaptation of material in *Diagnostic Standards and Classification of Tuberculosis* (1950) of the National Tuberculosis Association and in part a quotation from that text This material as adapted here is used by permission of the Association

The diagnosis of tuberculosis includes a determination of the presence and situation of a tuberculous lesion and an evaluation of its pathologic characteristics that is whether the lesion is open or closed Open lesions disseminate tubercle bacilli and therefore are dangerous or potentially dangerous to the patient and those with whom contact is made

Because active open pulmonary tuberculosis often exists without symptoms or abnormal findings both pulmonary tuberculosis with symptoms and pulmonary tuberculosis without symptoms are considered

Pulmonary Tuberculosis Without Symptoms The repeated roentgenologic examinations of apparently healthy contacts of known cases of tuberculosis disclose many asymptomatic but actually open and potentially dangerous lesions particularly in adolescents The mass roentgenologic examinations of apparently healthy groups in occupations or at ages in which tuberculosis mortality rates are relatively high have revealed lesions which may prove on further investigation and study to be minimal in extent but amenable to treatment

Further study and observation of these minimal lesions consist of history physical examination and serial roentgenologic examinations made at regular intervals of one to two months to determine the degree of instability of lesions Sputum studies are made from time to time temperature records are kept until it can be determined whether a lesion is active and in need of treatment Reactors to tuberculin should be examined roentgenologically each six months to one year

When applied to adolescents and adults mass roentgenologic examination of the lungs has as its primary object the detection of active or potentially active tuberculosis Mass tuberculin testing is commonly restricted to those of elementary school age or less although in rural communities and other low incidence areas it may be used to advantage among older age groups as a preliminary screening method

Pulmonary Tuberculosis With Symptoms Tuberculosis in its early stages seldom produces symptoms pronounced enough to cause the patient to consult a physician When a patient is unwell enough to consult a physician there is constantly a history of pleurisy poor recovery from respiratory diseases chest pain dyspnea hoarseness cough and hemoptysis The history of lassitude loss of appetite loss of weight and slight elevation of temperature and night sweats occurring in a young individual is particularly significant A physical examination may reveal moist rales over a limited area of the upper third of one side of the chest Occasionally the

abnormal signs are elicited over only a lower lobe Such findings require a roentgenogram to be made of the chest and if there is sputum it is examined for acid fast bacilli If negative on direct smear specimens should be concentrated and if necessary culture or guinea pig inoculation is made If there is no sputum gastric contents are obtained while the patient is fasting and are cultured or inoculated into guinea pigs The tuberculin test is given at the time of the first visit so that readings of the results may be made two or three days later A negative reaction to a tuberculin test is strong evidence that the physical findings are not due to tuberculosis A tuberculin reaction in itself should not be regarded as an indication of dangerous tuberculous infection except in infants

Obviously both the tuberculin test and the roentgenogram are of value in differential diagnosis of pulmonary disease Occasionally repeated stereoscopic films and examinations in the lateral oblique and lordotic positions may be of definite value

TUBERCULOUS COMPLICATIONS Three major complications of pulmonary tuberculosis are those in the larynx the trachea and the bronchi The larynx can be viewed readily The trachea and bronchi require bronchoscopic examination and sometimes even *this will not lead to the diagnosis if the lesions are beyond the bronchoscopist s* limited scope of vision Tracheal or bronchial lesions may give no symptoms and yet produce positive sputum Lesions of the trachea rarely cause wheezing The main clinical symptoms of bronchial disease are wheezing severe coughing and otherwise unexplained dyspnea all of which are the direct result of reduction of the lumen of the bronchus Massive collapse of a segment of a lobe of a lobe or an entire lung sometimes results from this interference with aeration and drainage The progress of this complication may seriously affect the management of a parenchymal pulmonary lesion

Involvement of the pleura is practically a constant finding in well-developed pulmonary tuberculosis Pleural effusions in young patients should be considered as of tuberculous origin unless proved otherwise A diagnostic tap for laboratory studies of the fluid is usually indicated Often at the time a patient complains of pain in the chest immediate fluoroscopic or roentgenologic examination mav not reveal evidence of effusion Repeating the examination after the pain has subsided will frequently result in the discovery of a pleural effusion

DIFFERENTIAL DIAGNOSIS OF PULMONARY TUBERCULOSIS The commoner conditions to be differentiated from pulmonary tuberculosis are the pneumonias pulmonary abscess low grade bronchiolitis secondary to chronic sinusitis bronchiectasis neoplasms (especially bronchogenic carcinoma) various pulmonary fibroses (especially silicosis) sarcoidosis and fungous infections

Among the commoner of the fungous infections are actinomycosis blastomycosis coccidioidomycosis and histoplasmosis Coccidioidomycosis is most often seen in the Southwest and strongly resembles tuberculosis in that it often gives rise to thin walled cavities and exudative lesions The symptoms are those of a prolonged influenza and the diagnosis rests on the recovery of *Coccidioides immitis* from the sputum or pleural fluid Histoplasmosis is found chiefly in the Middle Western and Southeastern states and it is not uncommon in these areas to see a number of small calcifications in roentgenograms of patients who have negative tuberculin and positive histoplasmin tests

In sarcoidosis patients often fail to react to the tuberculin test and have few if any symptoms in spite of roentgenograms showing hilar adenopathy and pulmonary lesions like those of far advanced tuberculosis miliary tuberculosis or silicosis

Roentgenologic shadows are never pathognomonic and when a person is in the age period in which cancer is most frequent and specific evidence of tuberculosis or other disease has not been obtained examination for malignant tumor should always be made

SCREENING CLASSIFICATIONS USED IN MASS ROENTGENOLOGIC SURVEYS During the years the public has become conscious of the ravages of tuberculosis and has done much through sanitary methods in the home and by demanding purer milk supplies to control this disease Those in charge of mass roentgenography seem to think that this method has been successful in the finding of many who have pulmonary lesions requiring further study

As the use of this technic increases the need for uniform standards and classification of screening film findings becomes apparent The systematic terminology used by the National Tuberculosis Association which has proved to be practical and informative is presented here

CLASSIFICATION OF PULMONARY TUBERCULOSIS *

Extent of Pulmonary Lesions *Minimal* Slight lesions without demonstrable excavation confined to a small part of one or both lungs The total extent of the lesions regardless of distribution shall not exceed the equivalent of the volume of lung tissue which lies above the second chondrosternal junction and the spine of the fourth or body of the fifth thoracic vertebra on one side

Moderately Advanced One or both lungs may be involved but the total extent of the lesions shall not exceed the following limits

Slight disseminated lesions which may extend through not more than the volume of one lung or the equivalent in both lungs

Dense and confluent lesions which may extend through not more than the equivalent of one third the volume of one lung

Total diameter of cavities less than 4 cm

Far Advanced Lesions more extensive than moderately advanced

Extent of Pulmonary Lesions Following Therapy In the case of temporary collapse the extent of disease existing immediately before the collapse shall be continued in the classification until re expansion permits reclassification

In the case of permanent collapse or pulmonary excision the extent of disease in a lung existing immediately before excision or collapse shall be continued in this classification through life except that such extension or new deposits of disease as appear any time postoperatively in this or the contralateral lung shall be additions to the preoperative classification The form site and duration of the procedure shall be added in parenthesis to the classification of extent of disease

In other cases the classification of extent will be changed as the change in the extent of the demonstrable lesion warrants Once the diagnosis of pulmonary tuberculosis has been established however the extent of the lesion cannot be classified as less than minimal even when clearing on the roentgenogram appears to be complete

Table 9 1 Bronchopulmonary Segments

RIGHT LUNG		LEFT LUNG	
LOBES	SEGMENTS	LOBES	SEGMENTS
Upper	Apical Posterior Anterior	Upper Upper Division	 Apical Posterior Anterior
Middle	Lateral Medial	Lower (Lingular) Division	Superior Inferior
Lower	Superior Medial Basal Anterior Basal Lateral Basal Posterior Basal	Lower	Superior Anterior medial Basal Lateral Basal Posterior Basal

* From *Diagnostic Standards and Classification of Tuberculosis* 1950 Edition with some re arrangements of order of presentation

Location of Lesions It may be useful to classify the extent of lesions in each lung separately When this is done however the combined extent for both lungs should always be designated

The location of lesions may be further designated by bronchopulmonary seg ments The terminology and classification of Jackson and Huber * have been adopted upon the recommendation of the American Association for Thoracic Surgery

CLINICAL CLASSIFICATION

The following definitions subject to the interpretation of the physician apply regard le s of the type of treatment given Collapse therapy or pulmonary excision in effect at the time of classification will be indicated under extent of pulmonary lesions The status of activity of lesions from a roentgenologic symptomatic and laboratory standpoint is designated first the status of a patient in terms of exercise shall be designated next

Roentgenologic Symptomatic and Laboratory Status *Inactive* Lesions as observed in serial roentgenograms must be stable except for extremely slow shrinkage and there must be no roentgenologic evidence of cavity Symptoms of tuberculous origin must be absent Sputum if any must be found negative for tubercle bacilli repeatedly not only by concentration and microscopic examination but also by culture or animal inoculation When a patient is not raising sputum or when there is any question concern ing the authenticity or adequacy of expectorated sputum specimens the fasting gastric contents or pulmonary secretions which have been aspirated from the tracheobronchial tree should be examined by culture or animal inoculation

These conditions shall have existed at least six months The period of inactivity shall be designated if known for example Inactive (6 months) Inactive (2 years) et cetera

Arrested The symptomatic and roentgenologic requirements of this group are the same as for inactive but the laboratory requirements are different

When sputum specimens or gastric contents have been found negative by repeated microscopic examinations of concentrates but not by culture or animal inoculation such patients cannot be classified as inactive but must be classified as arrested

Patients may also be classified as arrested even though culture or animal inoculation may be positive and among many concentrated specimens of sputum examined an occasional positive is found microscopically

These conditions shall have existed at least three months The period of arrest shall be designated if known for example Arrested (6 months) Arrested (2 years) et cetera

Active Lesions as observed in serial roentgenograms are usually progressive or retrogressive but may be stationary Symptoms of tuberculous origin are commonly present but may be absent Sputum and gastric contents almost always contain tubercle bacilli although in some instances tubercle bacilli cannot be demonstrated even after repeated cultures and animal inoculations With rare exceptions the tuberculin test is positive

The period of activity shall be designated if known for example Active (17 months) Active (3 years) The designations Active Improved or Active Unimproved may be used after an adequate period of observation or treatment

Activity Undetermined When activity has not been determined from adequate roentgenologic and laboratory examinations the disease must be designated temporarily as Activity Undetermined If a provisional estimate of the probable clinical status is necessary for public health purposes the terms (a) Probably Active or (b) Probably Inactive should be used Every effort should be made to classify cases and to avoid this category

Exercise Status The exercise status of a patient shall be designated

I The patient is not ambulatory

II The patient has been ambulatory for less than one hour daily

III The patient has been ambulatory for one hour daily for a period of two months

* Jackson C L and Huber J F Correlated Applied Anatomy of the Bronchial Tree and Lungs With a System of Nomenclature Dis of Chest 9 319 1943 quoted by Gray Henry in Anatomy of the Human Body Ed 25 Philadelphia Lea & Febiger 1948 pp 1134 1135

SCREENING CLASSIFICATIONS USED IN MASS ROENTGENOLOGIC SURVEYS During the years the public has become conscious of the ravages of tuberculosis and has done much through sanitary methods in the home and by demanding purer milk supplies to control this disease Those in charge of mass roentgenography seem to think that this method has been successful in the finding of many who have pul monary lesions requiring further study

As the use of this technic increases the need for uniform standards and classification of screening film findings becomes apparent The systematic ter minology used by the National Tuberculosis Association which has proved to be practical and informative is presented here

CLASSIFICATION OF PULMONARY TUBERCULOSIS *

Extent of Pulmonary Lesions *Minimal* Slight lesions without demonstrable excavation confined to a small part of one or both lungs The total extent of the lesions regardless of distribution shall not exceed the equivalent of the volume of lung tissue which lies above the second chondrosternal junction and the spine of the fourth or body of the fifth thoracic vertebra on one side

Moderately Advanced One or both lungs may be involved but the total extent of the lesions shall not exceed the following limits

Slight disseminated lesions which may extend through not more than the volume of one lung or the equivalent in both lungs

Dense and confluent lesions which may extend through not more than the equivalent of one third the volume of one lung

Total diameter of cavities less than 4 cm

Far Advanced Lesions more extensive than moderately advanced

Extent of Pulmonary Lesions Following Therapy In the case of temporary collapse the extent of disease existing immediately before the collapse shall be continued in the classification until re expansion permits reclassification

In the case of permanent collapse or pulmonary excision the extent of disease in a lung existing immediately before excision or collapse shall be continued in this classifica tion through life except that such extension or new deposits of disease as appear any time postoperatively in this or the contralateral lung shall be additions to the preopera tive classification The form site and duration of the procedure shall be added in parenthesis to the classification of extent of disease

In other cases the classification of extent will be changed as the change in the extent of the demonstrable lesion warrants Once the diagnosis of pulmonary tuberculosis has been established however the extent of the lesion cannot be classified as less than minimal even when clearing on the roentgenogram appears to be complete

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* From *Diagnostic Standards and Classification of Tuberculo is* 1950 Edition with some re arrangements of order of presentation

upon two factors (1) the annual decrease of reactors to tuberculin among various age groups in the population (2) the number of new cases and deaths reported annually that occur in families in which no tuberculosis has been known to exist previously Careful observance of these two factors will give the best indication of the proper time for re survey of any community or population group

DEMONSTRATION OF TUBERCLE BACILLI

The demonstration of virulent tubercle bacilli in the pulmonary secretions or gastric contents of patients with pulmonary disease is the one inescapable criterion of active pulmonary tuberculosis The results of other technical procedures including roent genologic examination may be equivocal However if concentrated specimens from a patient are examined for tubercle bacilli by an expert at frequent intervals the results whether positive or negative are of the greatest significance Since so much may depend upon a single sputum examination certain precautions are necessary to make the examination as reliable as possible

Preparation of Glassware All glassware used in sputum [collections and] studies must be scrupulously cleaned to ensure removal of any acid fast matter which may adhere from previous use Sputum bottles after being autoclaved to ensure sterility are held individually in a rapid stream of tap water to rinse out most of the organic matter The bottles are then boiled in soapy water for an hour after which they are placed in cleaning solution for at least 12 hours to destroy any remaining organic matter The cleaning solution is prepared by stirring 50 gm of technical grade sodium dichromate into one liter of concentrated sulphuric acid (sp gr 1 82) After removal from this solu tion the bottles are placed in a basin into which tap water is allowed to run rapidly for 30 minutes Finally each bottle is rinsed individually and dried and sterilized in an oven

Collection of Specimens *Sputum* A wide mouthed bottle of 30 to 300 ml capacity cleaned as indicated above should be used The bottle may have either a screw top or a snap cover The patient should be instructed to collect only secretion coming up the tracheobronchial tree from below the larynx i e not saliva or postnasal discharge As used subsequently the term sputum will refer only to secretion of this type coming from the lungs

Gastric Contents In certain cases examination of the gastric contents for tubercle bacilli is essential This is particularly true in children or other patients who do not raise sputum as well as in patients whose sputum has failed to show tubercle bacilli after repeated examinations The technique of gastric lavage is as follows The material is collected after at least an 8 hour fast A Levine tube carefully cleaned sterilized and lubricated with sterile water is introduced through one nostril the patient is asked to swallow as soon as he feels the tip of the tube in his throat With a glass syringe of 20 to 50 ml capacity the gastric contents are withdrawn and placed in a sterile 250 ml Erlenmeyer flask The patient is then given 30 to 50 ml of sterile water to drink and the gastric contents are again aspirated and added to the first aspiration The specimen must be sent to the laboratory immediately and must receive the attention of the tech nician at once Prolonged standing may make the recovery of tubercle bacilli difficult or impossible Gastric specimens which must be sent through the mails may be frozen and shipped in dry ice It should be realized that in the case of ambulant patients a walk or long ride to the doctor s office may cause the stomach to empty before a gastric specimen is obtained

Other Clinical Specimens For the demonstration of tubercle bacilli in other body fluids such as pleural pericardial or peritoneal effusions spinal fluid or pus from cold abscesses or sinuses essentially the same methods are used as for sputum

To recover tubercle bacilli from urine [at least] 1 000 ml taken from a well mixed 24-hour specimen [is required]

Positive results from such voided specimens should be confirmed with catheterized specimens These may be treated in the manner given above Also it is advisable to inoculate the untreated sediment into guinea pigs

In the examination of feces [at least 5 gm is required]

Negative Sputum If sputums and gastric contents are carefully and repeatedly examined as outlined above negative results are of distinct diagnostic value However it should be emphasized that a given specimen can never be called Negative for tubercle

IV The patient has been ambulatory for at least two hours daily for a period of at least two months

V The patient is living under ordinary conditions of life

The exercise status of a patient shall be designated following the designation of the roentgenologic symptomatic and laboratory status for example Inactive (6 months) III

Classification Classifications of roentgenologic findings can be stated comparatively easily In summary they may be listed as follows

Screening Films of Any Size

1 Essentially negative
2 Unsatisfactory (technic)
3 Calcification only
4 Suggestive of tuberculosis
   a Minimal
   b Moderately advanced
   c Far advanced

   Note *If a provisional estimate is necessary the terms probably active or probably inactive should be used for public health purposes*
5 Suspected tuberculosis
6 Probably nontuberculous
   a Cardiac changes
   b Pleural changes
   c Other changes
7 Diagnosis deferred

Explanation of Classification of Impression from Survey Films Following is a brief explanation of these classifications

1 Essentially negative All apparently normal chests without evidence of present or past disease and without calcification

2 *Unsatisfactory* Technic unsatisfactory for screening requiring retake

3 Calcification only (distinctly present in parenchyma or in hilum without infiltration)

4 Suggestive of tuberculosis Mottled strand like shadows especially in upper lobes with or without evidence of cavity or associated calcification The location and extent of abnormal shadows must be taken into consideration

a Minimal
b Moderately advanced
c *Far advanced*

5 Suspected tuberculosis Shadows of such character that opinion must be reserved until clinical studies can be made This category includes such conditions as suspected hilar lymphadenitis evidence of collapse therapy and pleural effusion

6 *Probably nontuberculous* a *Cardiac changes* Abnormalities of size shape or position of heart or aorta

b *Pleural changes* Localized or extensive *pleural thickening in the absence of* other evidence of tuberculosis

c Other changes such as tumors silicosis other pneumoconioses or pneumonia

7 Diagnosis deferred Abnormal shadows not classifiable in the categories described above

Films regarded as suggestive of tuberculosis should be further qualified as (1) probably active or (2) probably inactive However the diagnosis of tuberculosis and the estimate of clinical status of such cases should be provisional for all initial or screening examinations even if the initial survey film is a 14 x 17 inch celluloid film

It should be emphasized that final diagnosis should *never* be made from survey roentgenograms alone regardless of the technic employed One can be certain of exact diagnosis only after meticulous laboratory and clinical studies Such studies should include clinical history a physical examination sputum examinations gastric or tracheal cultures when necessary tuberculin testing and serial roentgenologic examination The above classification therefore is for screening purposes only

Intervals for Re Surveys No arbitrary time interval can be *recommended for* re survey of large population groups The completeness of the initial survey adequacy of isolation and treatment of newly discovered infectious persons and *follow up* examinations of contacts of known cases will determine the need for or frequency of re survey Epidemiologically the real test of the time interval of re surveys will be based

of tuberculosis are in the vast majority of instances highly sensitive to PPD or Old Tuberculin A dose of 0 00002 mg of PPD or 0 01 mg of Old Tuberculin is recommended for the initial dose for those living in areas having a high morbidity or mortality from tuberculosis for those with a history of severe reactions following previous administration of tuberculin for those with a history of intimate contact with persons with clinically manifest tuberculosis and for persons with extrapulmonary forms of this disease

If PPD is used a standard product which is available on the market is dissolved in the requisite amount of the diluent supplied so that each 0 1 ml of the dilution will contain 0 0001 mg of PPD To prepare the dose of 0 1 mg of Old Tuberculin recommended for case finding purposes 0 1 ml of concentrated Old Tuberculin is added to 9 9 ml of sterile physiological salt solution thus making a dilution of 1 100 One milliliter of the 1 100 dilution of Old Tuberculin is transferred to 9 0 ml of sterile physiological salt solution thus making a dilution of 1 1 000 Each 0 1 ml of this dilution will contain 0 1 mg of Old Tuberculin Purified Protein Derivative and the diluted Old Tuberculin are best prepared fresh Solutions more than a few days old should not be used When not in use diluted solutions should be kept in a refrigerator

For the present with certain exceptions as noted the use of a single dose is recommended Further studies on the specificity of the larger doses of both PPD and Old Tuberculin are being made

The intracutaneous tuberculin test (Mantoux) with either PPD or Old Tuberculin should be read 48 or 72 hours after the injection Readings should be made in a good light with the arm slightly flexed Response to injection is classified as positive negative or doubtful Reactions may be classified arbitrarily as one two three or four plus depending upon the extent of induration measured at its widest diameter A reaction showing some definite induration more than 5 mm and not exceeding 10 mm in diameter is recorded as a one plus (+) reaction A two plus (++) reaction is an area of induration measuring from 10 to 20 mm in diameter A three plus (+++) reaction is characterized by marked redness and induration exceeding 20 mm in diameter A four plus (++++) reaction consists of severe induration and an area of necrosis A reaction with a trace of induration measuring 5 mm or less in diameter is rated as doubtful Redness without associated induration does not constitute a reaction

Syringes which have been used for the administration of PPD or Old Tuberculin should not be used for the administration of preparations such as coccidioidin histoplasmin or other diagnostic reagents since PPD and Old Tuberculin are difficult to remove from the syringes

Scarification or Pirquet Test This procedure is carried out by applying a drop of undiluted Old Tuberculin to the cleansed skin preferably over the region of the deltoid muscle and scarifying the skin through the drop of tuberculin by means of a sterile needle or small borer Care should be taken that the scarification does not cause bleeding The tuberculin should be permitted to remain in situ for about 20 to 30 minutes before the excess is removed The results of the test are read 48 to 72 hours later and are interpreted in the same manner as the intracutaneous test If there is no tuberculin reaction by this method it is advised that it be followed by the administration of 0 0001 mg of PPD or 0 1 mg of Old Tuberculin using the intracutaneous method

Patch or Percutaneous Test of Vollmer The patch test is carried out by applying a prepared patch to the skin in the region of the sternum or between the scapulae The patch is composed of two squares impregnated with concentrated tuberculin prepared from cultures grown on a synthetic medium and one control square impregnated with the concentrated culture medium The skin is first cleansed with acetone and the patch applied and permitted to remain in situ for 48 hours The reaction is read 48 hours after the removal of the patch A reaction appears as an inflamed area of varying intensity In those highly sensitive to tuberculin it is advisable to remove the patch as soon as the person complains of irritation since a severe local reaction with blistering may occur If the patch test is negative 0 0001 mg of PPD or 0 1 mg of Old Tuberculin should be injected

Of the procedures here described the intracutaneous method is the most sensitive and permits the administration of graduated and accurate amounts thus making it

bacilli until it has been examined by adequate cultural methods and animal inoculation as well as by examination of the direct and concentrated smears In a patient with a demonstrable parenchymal infiltration in whom tubercle bacilli cannot be demonstrated causes for the lesion other than active pulmonary tuberculosis should be sought

In a tuberculous patient no specified number of negative sputum examinations gives absolute assurance that another specimen may not show tubercle bacilli Nevertheless it is convenient to designate an *arbitrary number of negative examinations which may* be taken as presumptive evidence of bacteriologic remission To designate sputum as negative in this limited sense it is recommended that three adequate specimens of sputum or of gastric contents taken at least a week apart be found negative by culture or by animal inoculation

TECHNICAL PROCEDURES

The Tuberculin Test The tuberculin reaction is an acute local specific inflammation resulting from the injection or application of tuberculin Few procedures employed in the control of tuberculosis are of greater value than the tuberculin test As an index of the extent of infection in communities it is of the utmost epidemiological value It locates sources of contagion and provides a guide for case finding and family and group supervision Its importance in the diagnosis of tuberculosis rises constantly as the rate of infection in the population decreases and as the total number of cases of active disease declines Whereas its value in clinical diagnosis once was exclusive rather than definitive in character the tuberculin test is becoming a more positive indication of significant infection Finally with the expansion of BCG vaccination programs and the importance of determining the relation of artificial infection with an organism of low virulence to the development of individual resistance the tuberculin test has become of special significance as an indication of the take of the vaccine

Obviously once BCG has been administered the tuberculin test loses its specific value as an indicator of natural infection

A reaction to certain specified amounts of tuberculin is an indication of tuberculous infection With few exceptions definite sensitivity to tuberculin once acquired persists through life This sensitivity may vary in intensity and may temporarily decrease or disappear in the course of high fever exanthematous disease miliary tuberculosis and the last stages of pulmonary tuberculosis A very small percentage of persons may fail to react to tuberculin even after infection resulting from exposure to cases of open tuberculosis or after the administration of vaccine prepared from living or dead tubercle bacilli The tuberculin test remains negative in a high percentage of cases of sarcoidosis

At the present time Purified Protein Derivative (PPD) and Old Tuberculin (OT) are widely used Purified Protein Derivative (PPD) is the tuberculin of choice and is recommended as the standard for comparative studies It is prepared by growing tubercle bacilli on a synthetic medium of known composition The tuberculin protein produced in the medium is then obtained by precipitation with ammonium sulfate at neutrality and further purification Old Tuberculin is prepared by concentrating by heat the glycerine broth medium upon which the tubercle bacillus has grown

TECHNIC The method of choice in administering tuberculin and the one recommended for accuracy and for all general purposes is the intracutaneous (Mantoux) test Occasionally however special circumstances may make other methods of administration preferable In the following paragraphs the intracutaneous method is described as the standard procedure A description is also given however of the scarification (Pirquet) test and of the Vollmer patch test which is frequently employed when there is objection to the use of a needle Neither of these tests is considered as accurate as the intracutaneous test in which a measured amount of tuberculin is introduced into the skin

Intracutaneous or Mantoux Test The intracutaneous tuberculin test is best carried out by injecting the desired concentration of PPD or Old Tuberculin into the cleansed skin of the forearm This injection is made with a short (half inch) sharply beveled 24 or 25 gauge platinum or steel needle and a tuberculin syringe *When the injection* is properly made a wheal should appear immediately at the site of injection

The use of a single dose of 0 0001 mg of Purified Protein Derivative (PPD) or 0 1 mg of Old Tuberculin is recommended for case finding programs This dose is recommended since persons showing roentgenographically demonstrable lesions characteristic

diagnosis of such diseases must depend on laboratory studies of materials obtained from patients including direct microscopic examination of sputum cultures of sputum pathogenicity tests of organisms isolated and sensitivity and serologic tests *Actinomyces bovis Nocardia asteroides Candida albicans Geotrichum candidum* and *Histoplasma capsulatum* are best detected in stained smears of the sputum (see Chapter 17)

India ink preparations are used to demonstrate the capsule which surrounds the budding yeastlike cells of *Cryptococcus neoformans* A medium containing penicillin and streptomycin supports the growth of pathogenic fungi with two exceptions *Actinomyces bovis* and *Nocardia asteroides* which are susceptible to penicillin and streptomycin and must be cultivated on other mediums Most fungi known to cause pulmonary infections are pathogenic for one or more laboratory animals such as mice rabbits and guinea pigs Many patients with fungous infections manifest sensitivity to the invading organism or its products and intradermal injection of an appropriate antigen results in a tuberculin type delayed reaction The interpretation of the positive result is the same as that applied to the tuberculin test The reaction becomes negative in the acutely ill patient as a result of anergy A positive serologic reaction in coccidioidomycosis histoplasmosis and blastomycosis in which the complement fixation tests have been used most extensively is indicative of an active progressive disseminated infection Such a test is therefore diagnostic Cross reactions are known to occur between *Blastomyces* antigens and antiserum against *Histoplasma capsulatum* Any serum tested should be tested against both *Blastomyces* and *Histoplasma* antigens

TUBERCULIN COCCIDIOIDIN AND HISTOPLASMIN SENSITIVITY IN RELATION TO PULMONARY CALCIFICATIONS In view of the evidence that histoplasmosis and coccidioidomycosis may be etiologically associated with pulmonary calcification Beadenkopf and associates included these antigens among the skin tests given as part of the tuberculosis control program at the University of Chicago Only 30 per cent of the students enrolled came from Chicago and vicinity the greater number came from practically all parts of the United States and several hundred were from foreign countries The report was based on the study of 6 000 students registered at the University from January 1 to October 1947 This study on a population of diverse origin confirms the results of other recent surveys It was found that (1) the prevalence of tuberculin sensitivity progressively increased from 10 per cent among the youngest age group to 57 per cent among those 50 years of age (2) the rate of histoplasmin sensitivity began at 10 per cent among the youngest group and reached 38 per cent in the oldest group (3) pulmonary calcifications were associated with histoplasmin sensitivity twice as frequently as they were with tuberculin sensitivity and (4) the prevalence of both histoplasmin sensitivity and pulmonary calcifications was highest in the Lower Mississippi Basin of the United States

In young people living in or adjacent to the Mississippi River Basin and the bordering states of the United States there is a prevalence of pulmonary calcification far in excess of that which can be explained on the basis of tuberculosis as revealed by tuberculin cutaneous testing In this area from one third to four fifths of young persons have pulmonary calcification Only one half or less of those who have pulmonary calcification have positive tuberculin reactions

Anergy or loss of sensitivity to tuberculin from complete healing of the tuberculous process may be an acceptable explanation in some instances but this cannot explain the decided prevalence of pulmonary calcification in this area

Cox and Smith Aronson Saylor and Parr and others have shown that in certain areas of the West coccidioidomycosis is responsible for a primary complex which goes on to calcification The suggestion of Smith that histoplasmosis occurs most frequently in the area where pulmonary calcification is high in those whose reaction to tuberculin is negative seemed to offer a good working hypothesis

possible to detect cases with a low threshold of sensitivity to tuberculin The general use of the Pirquet and the patch test is not recommended since they are not so efficient as the Mantoux test

Methods of Case Finding in Tuberculosis The facilities used in case finding have continually been improved At first the only means of finding tuberculosis was by physical examination then came the discovery of the bacilli and the use of the microscope and then the roentgen rays Following this the tuberculin test was applied with much success and finally the miniature roentgenogram was developed

The mass roentgenologic survey method in large communities is probably most effective and cheapest this is true especially of the 70 mm machine which may be used with all the facility of the earlier developed 35 mm and yet possesses most of the advantages of the larger (14 by 17 inch) films The cost is not large and the results are probably of more direct value than those obtained with any other method The roentgenologic method finds the significant lesions which can be quickly treated

Several things roentgenologic surveys do not do Since roentgen shadows are not specific they are not pathognomonic of tuberculosis Subsequent sputum analyses and tuberculin tests are necessary in 3 to 5 per cent of all cases surveyed often revealing that conditions which at first were called tuberculosis actually are some other disease Second the roentgen rays will not always reveal the presence of tuberculous lesions since about 30 per cent of the chest area is hidden and since many early lesions do not cast shadows

The tuberculin test method as introduced by Opie Long Meyers and a large number of others has many advantages not possessed by any other technic A tuberculin test helps to rule out nontuberculous disease When the reaction is positive in infants and young children and even in young adults the test is a certain indication of proximity to open tuberculosis Because more persons will reach adult life in the future without infection the test will become increasingly useful

One of the greatest benefits of tuberculin testing in survey work is found in communities where only 50 per cent or less of the population has a positive tuberculin reaction Obviously the majority of the roentgenograms of those with negative tuberculin reactions would be wasted The tuberculin method in such circumstances should be used first and followed by roentgenograms of the positive reactors Finally when used fully the tuberculin test is perhaps the best index of the efficiency of a tuberculosis control program in any community

The tuberculin survey method is the most practical in communities where the infection rate is low Positive reactors may be given roentgen examinations after a tuberculin survey on entering and on leaving school Follow up of all contacts should be made and health education intensified

In larger communities where the infection rate is high probably the best method is to give roentgen examinations to everybody more than 15 years of age and the tuberculin test to all those with suspected lesions Although a few small lesions will escape the roentgenologic survey the number will not be of great importance The cost of each method when used in its respective proper setting is probably about the same based on the total cases surveyed The roentgenogram is cheaper per number of cases found and number of deaths from tuberculosis while the tuberculin test method is more useful in small communities and is above all a sound index of the control of the disease These two principal methods should and do supplement each other the choice and sequence depending on time place and available facilities

Differentiation of the Active Phases of Pulmonary Mycosis and Tuberculosis *Actinomyces bovis Nocardia asteroides Coccidioides immitis Blastomyces dermatitidis Blastomyces brasiliensis Histoplasma capsulatum Cryptococcus neoformans Candida albicans* and *Geotrichum candidum* cause subacute and chronic infections of the lungs which resemble tuberculosis These diseases often cannot be separated and never can be definitely diagnosed on clinical findings alone The incidence of these mycotic infections is as a rule low in comparison with the incidence of tuberculosis In the absence of positive smears, cultures and guinea pig inoculations for tuberculosis these mycotic diseases should be considered The

it is certain that no outside contact has been made within a year there has never been during the residence of foreign observers in these communities the slightest evidence of acute infections of the respiratory tract Let there be contact with out siders and these communities will have an epidemic of colds within 2 days It must be emphasized however that despite the studies which have been made in these communities in reality the number of studies has been small and the time spent in these localities by investigators has been for practical purposes too short to estab lish the conclusions which have been reached

From the data now obtainable it seems likely that a virus is present in the nasal discharge of patients during the first 3 or 4 days of an attack of a viral cold This virus when dispersed in the air or on objects in the open air remains infective for about 5 hours The virus is said too to be spread by contaminated hands either directly or indirectly Likewise drinking glasses cups forks and spoons are excellent vehicles for the transmission of the cold virus as well as other germs from one person to another However it has not been proved that colds are thus epidemically disseminated

INCIDENCE Colds are more prevalent during the winter months than in other seasons in both *cold and temperate climates* They seem to go around several times during the winter months of each year

The prevalence of colds is about the same in the agreeable climates and the disagreeable climates Sleeping on sleeping porches in well ventilated bedrooms or in poorly ventilated bedrooms does not affect individual susceptibility

During the winter months of each year there is a tendency for colds to increase when there is a decrease in temperature below the normal range of temperature for the corresponding week and also for the preceding week

ETIOLOGY Among the various etiologic agents which deserve mention are viruses bacteria chilling physical agents allergies and climate

Viruses Dochez and his associates concluded that colds can be transmitted from man to the chimpanzee and from man to man by means of Berkefeld filtrates of nasal washings from individuals suffering from acute colds and that the cause of colds is a filtrable agent which in all likelihood belongs to the group of so called submicroscopic viruses The conclusion seems justified that a filtrable virus is the cause of certain colds and probably of some epidemics but not all of them

Bacteria Colds of bacterial origin begin with a severe sore throat with the nasal discharge and congestion appearing later as the infection extends upward into the nasal passages It seems that bacterial colds may appear in the course of a viral cold and are probably due to secondary bacterial invaders The vasomotor reaction may so disturb the equilibrium between the host and the bacteria of the nose as to favor the development of infection

Allergies The symptoms which allergens produce in the upper portion of the re spiratory tract are essentially those of an acute coryza However there seem to be regularly each year the so called spring and fall colds in addition to these allergic phenomena

Physical Agents These include climates good or bad chilling of the body and injury of the mucous membranes by particulate matter

The exposure of an individual to drafts and *chilling of the whole or of portions of the body* may result in the symptoms of a cold It has been demonstrated that chilling of the body surfaces leads to peripheral vasoconstriction with attendant peripheral stasis and anoxemia to lowered leukocyte response and to impairment of the phagocytic capabilities of the fixed tissue cells including those of the nasal mucous membrane Chilling of the body surfaces causes a fall in the temperature of the nasal and pharyngeal mucous membranes and this chilling and the accompanying congestion increase sus ceptibility to a cold

The occurrence of colds in different geographic locations has illustrated that the attack rates are uniform irrespective of latitude longitude or climate The period of high incidence and the occurrence of epidemics in widely different localities often take

Christie and Peterson accordingly began to perform cutaneous tests with extracts of *Histoplasma capsulatum* and to look for nonfatal examples of the infection The fungus *Histoplasma capsulatum* had been reported to cause acute and subacute granulomatous lesions progressing with great regularity to a fatal termination Their studies enabled them to observe examples of a benign form of histoplasmosis and to discover that many people react to extracts of the fungus

The studies of Christie and Peterson and of Palmer and Furcolow show a remarkable relationship between sensitivity to histoplasmin and the development of pulmonary calcification The early development of sensitivity to histoplasmin followed by the development of the calcification at a rate much greater than the age specific rate of sensitivity to tuberculin suggests that most of the lesions are more likely to be related to the development of sensitivity to histoplasmin than to be associated with tuberculosis

According to Aronson and his co workers Indians of the United States and Alaska who failed to react to the intracutaneous injection of 0 00002 and 0 005 mg of Purified Protein Derivative of tuberculin revealed calcified pulmonary nodules Among 704 children living on the Pima Agency who did not react to tuberculin calcified pulmonary nodules were observed in 102 among 419 on the Wind River Agency only 1 of those who failed to react to tuberculin had a calcified pulmonary nodule among 332 children living on the Turtle Mountain Agency who did not react to tuberculin 15 had calcified pulmonary nodules and among the 592 children of the Rosebud Agency and the 977 children living in southeastern Alaska who failed to react to tuberculin 6 and 2 presented calcified pulmonary nodules Because of the high incidence of calcified pulmonary nodules in tuberculin negative children living on the Pima Agency the possibility that the nodules were coccidioidal was investigated A large percentage of children of school age living on the Pima San Carlos and Sells agencies and a small percentage of those living on the Fort Apache Agency reacted to the injection of 0 1 ml of a 1 1 000 dilution of coccidioidin

DISEASES OF THE RESPIRATORY PASSAGES AND THE LUNGS DUE TO FILTRABLE VIRUSES

The Common Cold The consensus is that the common cold and its complications in the United States are the cause of more illness and disability than any other illness It must be clear that the term cold is applied by the physician and laity alike to various acute subacute and even chronic maladies of both the upper and lower portions of the respiratory tract and thus statistical data concerning the incidence and the duration of the common cold are well nigh impossible to interpret The term coryza refers to a cold in the head

An acute respiratory infection often does not remain limited to any one portion of the respiratory tract For instance rhinitis today may become sinusitis pharyngitis laryngitis tracheitis or bronchitis tomorrow In many instances the sequence may be changed partially at least Tracheitis laryngitis or pharyngitis may be largely over in a day or two to be followed by rhinitis In reality many individual regularly with each cold have symptoms which commence as pharyngitis laryngitis or tracheitis to be followed by coryza

EPIDEMIOLOGY Colds are cosmopolitan in distribution in regard to geography altitude and age incidence The attack rate is highest and the complications are most frequent and most serious in infants and in the aged From these ages the rate declines to a low point in the age group 15 to 24 years rises from 25 to 34 years again declines until it reaches a minimum in the age group of 50 to 60 or 65 years and then tends to increase

ISOLATION Some extraordinarily interesting observations on the occurrence of colds have been made in isolated communities In the arctic and polar regions when

respiratory tract such as laryngitis tracheitis bronchitis and pneumonia Closely allied to colds are the diseases influenza and tonsillitis

It is postulated that complications of the common cold may occur when drain age from the sinuses or middle ear is obstructed by swelling of the mucous mem branes The presence of virulent organisms in the nose and throat at the time of the cold is likely to originate disease when the subject is in poor health

DIAGNOSIS The diagnosis of a common cold is a presumptive one until the patient has fully recovered The diagnosis of a complication of a bad cold depends on the excessive localization of symptoms and signs to the sinuses ears or lungs It is well to recall that some of the aches in the back muscles and head are due to fever Anorexia is due to loss of the senses of smell and taste Constipation is occa sioned by lack of food In some instances diarrhea may be due to cold remedies which have been self administered

Influenza Influenza is known also as grippe grip febrile catarrh catarrhal fever acute nasopharyngitis epidemic catarrh and epidemic influenza

Influenza is an acute self limited infectious disease of man which is caused by a virus of the influenza group The illness is characterized by symptoms which are predominantly constitutional although the infection is limited to the respiratory tract It tends to occur in epidemic forms Two specific and distinct etiologic types of the disease are now recognized one termed influenza A is caused by infection with influenza A virus the other termed influenza B is caused by infection with influenza B virus (Horsfall 1940) The cause of pandemics of influenza has not been established

Among the periodic outbreaks of influenza the major ones are apparently due chiefly to influenza A virus others may be due wholly or partly to influenza B virus still other viruses may produce morbidity but may fail to show signs of their pres ence although the illnesses produced by these as yet unproved viruses are clinically like those of known influenza A and B viruses Influenza A virus varies from one outbreak to another antigenically and in its rapidity of spread in the population Influenza B virus produces epidemics of limited scope Influenza A virus usually lies quiescent for 21 months out of 24 During this quiescent interval there is some relationship between the individual immunity and the level of neutralizing antibodies in the serum

Influenza viruses may vary greatly from one epidemic to another As an example of the variability of the influenza virus according to Tamm Kilbourne and Horsfall the influenza B virus isolated in the New York area during the 1950 epidemic was so dif ferent from previous strains as to throw doubt on the desirability of continuing the use of a routine strain exclusively in prophylactic immunization Using the chick embryo technic four strains of influenza B virus were isolated from hospitalized patients Tamm Kilbourne and Horsfall had for comparison the routine virus and five strains isolated by the same technic during previous epidemics (1945 1947 1949) Thus it is possible that it will be necessary to use a polyvalent influenza B vaccine in the future rather than to depend on the Lee strain exclusively

Influenza viruses are not known to be related to the etiology of the common cold

EPIDEMIOLOGY The consensus is that a primary epidemic occurring in any locality after a period of years nearly free from the disease is a uniform disease Influenzal epidemics are independent of meteorologic and climatic conditions

The number of instances of the disease increases rapidly to a maximum and then decreases to the endemic level so that the graph of the curve is almost symmetric The fatality rate although variable is low

Primary epidemics are followed by secondary epidemics which are less extensive and more slowly reach their maximum from which they decline more slowly and irregularly Because of secondary pulmonary complications the fatality rate is greater in the secondary epidemics than in the primary epidemic

Influenza regularly occurs in endemic or seasonal form in most countries every

place synchronously too fast for human travel and thus transport of the offending etiologic agent

Temporary irritation or injury of the membranes of the nose and throat by *particulate matter* such as various dusts will cause the nasal symptoms characteristic of colds If the exposure to irritation is prolonged the symptoms may subside or may become chronic and severe Smoking of cigarettes may be a factor in keeping up a low grade chronic irritation Irritating chemical gases or air filled with particulate matter (dusts) or overheated dry air predisposes to colds

Indeterminate differences in individual susceptibility to colds cannot be explained

Some individuals seem to have hypersusceptibility to the common cold Others are rarely susceptible to colds under any circumstances These variations cannot be adequately explained by hypersensitivity due to an allergic state or by immunity

In persons who have frequent colds the vasomotor response of the nasal mucous membranes to peripheral cold stimuli is much less in degree and slower in appearance than in normal persons and during an acute cold this reaction is entirely absent

Persons who are taking active physical exercise and who have accustomed themselves to rapid changes in temperature are less susceptible to colds than other persons even if they have the foredescribed mucous membrane attributes

SYMPTOMS The common cold commences in one or more of several ways Often *the first symptom is a feeling of congestion in the nose with sneezing Other colds* will begin first with a general ache and perhaps slight fever the temperature from 99 to 100 F (37 2 to 37 7 C) and still other colds will be accompanied with general aching malaise and loss of appetite for a day or more before there are any respiratory symptoms Once any one or all of these discomforts are established symptoms in the nose and throat ensue There is localized or generalized burning in the nose and throat The discomforts spread and the mucous membranes of the nose nasal *sinuses and pharynx become red and swollen As the result of swelling of its mucous* membranes the nose becomes obstructed so that mouth breathing is more comfortable than nose breathing If the sinuses are prevented from draining by the swelling there is a dull ache in the face and head The inflamed mucosa secretes a watery serous exudate which drips almost constantly from the nose This watery secretion is irritating to the outside skin of the nose and the upper lip

The inspiration of cold air dust or tobacco smoke hurts the nasal passages *Often confirmed cigarette and cigar smokers* cannot tolerate their favorite pastime because the smoke hurts the nose and throat and may make the headache worse Involvement of the eustachian tubes causes noises created by the mucus and air in the tubes to be heard as popping and roaring sounds in the ears If the eustachian tubes become closed temporary deafness follows The throat becomes dry swollen and uncomfortable The tongue is dry The senses of taste and of smell are impaired or lost A decrease in the degree of the acuity or a permanent loss of either or both the senses of smell and taste may follow a cold However recovery of the senses (hearing smell and taste) is almost invariably the rule

Slight fever may accompany acute colds A moderate or a high fever the temperature more than 102 F (38 8 C) indicates more than a cold for instance, some *complication such as sinusitis otitis media tonsillitis bronchitis* or pneumonia

Involvement of the lower part of the respiratory tract with hoarseness and coughing is commonly present in some individuals in others it rarely occurs

EXAMINATION Examination by physical means reveals the linings of the nose the pharynx and throat to be reddened The voice often is husky or lost Nasal or labial herpes may be found If the mucous membranes of the tear ducts are swollen the eyes are watery and the conjunctivae are injected The lungs are clear

An important reason for a complete physical examination of the patient who has a bad cold is to search for evidence of complications The complications of colds are sinusitis otitis media and more than transient infections of various parts of the

pneumonia of influenza is generally combined with capillary bronchitis upon which it supervenes The instances in which pneumonia is combined with bronchitis are less commonly fatal than those in which capillary bronchitis alone existed

Gastrointestinal Form This form of influenza begins with severe colicky pains vomiting and diarrhea In this form and in the nervous form the catarrhal symptoms are often altogether absent or only come on late in the course of illness

Nervous Form The nervous form is characterized by intense headaches and backache The backache may be as severe as that which occurs during a hemolytic crisis or in smallpox The headache frontal in some cases occipital and occasionally vertical is often persistent continuing for some days after the patient s temperature has subsided to normal values

The nervous form of influenza occurring in tropical and subtropical regions may be difficult to differentiate from dengue (six day fever breakbone fever) except by laboratory tests

SYMPTOMS The onset of influenza is sudden On this point all of those who have reported the epidemics which have occurred from remote to recent times have agreed

Fever is constantly present except in cases in which pronounced nervous symp toms exist The temperature may reach high figures 105 8 F (41 C) early in the course of the disease but if defervescence is rapid no great harm is done The fever lasts from 3 to 10 days the longer the fever continues the more likely it is to resolve by lysis

Coryza or *rhinitis* or both vary greatly in different epidemics of influenza In most instances there is some degree of involvement of the respiratory system In the total number of persons who have the disease these symptoms are not severe The respiratory rate is usually not increased unless there are pulmonary complications An early increase in the respiratory rate is an ill omen

Cough is not always a prominent feature and often it is absent The cough bears but little relation to the extent of the physical signs in the lungs It is likely to occur in paroxysms which are sometimes very exhausting Toward the end of the grave illnesses cough which may have been very troublesome indeed in the earlier stages generally ceases even though the lungs are full of rhonchi and rales

Pain may be the earliest symptom of influenza Pains in the limbs and back shivering and often severe headache may be the only symptoms for a day and a night The pains are referred almost invariably to the muscles and soft tissues and to a less extent to the joints rather than to the bones themselves The commonest sites of pain are in the muscles of the thighs and calves the lumbar muscles and muscles of the back of the neck and to a less extent the knees and ankle joints The pains are severe but differ considerably from those of trench fever and smallpox In these diseases the pains are most frequently referred to the bones themselves particularly to the inner and outer edge of the tibiae and the bones are often very tender to palpation The acute nocturnal exacerbations of the pains are more marked in trench fever than in influenza

The abdominal pains in some of those who have influenza are more general and more frequently referred to the lower central part of the abdomen than are the pains of appendicitis There is little or no muscle guarding and the tenderness to deep palpation is not more marked in the right iliac region than in the left The pain does not tend to become localized and the fever and pulse rate do not indicate a progressive lesion

There are those who have occipital headache and pain in the neck closely resembling the pain of cerebrospinal meningitis but as a rule the rigidity of the neck is much less marked In some of these patients it is impossible to say that Kernig s sign is absent the phenomenon is more of the nature of a clasp knife rigidity than a true Kernig s sign Lumbar puncture may however be necessary to exclude cere

year The exact relationship of endemic influenza to the classic pandemic type of influenza is unknown Many believe that endemic influenza and the minor outbreaks are a repetition of the classic type but during a period of attenuation of the virulence of the virus others suggest that endemic or sporadic influenza is an infection of a different nature or type altogether

The frequency of the outbreaks of influenza and its tendency to reach pandemic proportions affecting equally those of all economic and social levels of society traveling from continent to continent at a speed equal to that of human travel affecting and destroying millions of lives (the 1918 1919 pandemic with 21 642 283 deaths) respecting no barriers set up for control by public health organizations and leaving behind ill health and disease stigmatize influenza as the greatest pestilence of our time The importance of this disease in addition to its own morbidity lies in its power to induce a condition of particular susceptibility to attack by secondary pathogenic organisms, particularly the hemolytic streptococcus and tubercle bacilli

The pneumonia which often follows influenzal infections is caused by an associated bacterial invasion of the lungs In cases of *Staphylococcus aureus* pneumonia complicating sporadic instances of influenza the staphylococcus is present in almost pure culture The hemolytic streptococcus (type A) is more commonly present in the various epidemics of pneumonia following influenza The pneumococci and *Hemophilus influenzae* may be the bacterial agents responsible for some of the pulmonary complications It seems reasonable that the influenza virus itself may be capable of originating bronchopulmonary reaction if not an actual pneumonia The diffuse bronchiolitis which may occur after epidemic influenza may represent a true viral lesion of the lung

The effect of endemic influenza on the morbidity as well as on the mortality rate from pneumonia and tuberculosis and on the birth rate is less marked than that of the pandemic disease Prevalence of endemic influenza is more influenced by seasonal conditions as compared with the pandemic type

PATHOLOGY *The evidence would indicate that the filter passing virus of influenza* gains access to the body by the respiratory tract The influenza virus injures or *destroys the ciliated epithelium lining the air passages* It inflicts an initial damage of the epithelium of the respiratory tract at special selective sites In this respect stress is laid on the importance of the postnasal area as an important primary focus *in the dissemination of* infection in the body From this area the virus may directly spread downward inflicting severe damage in the ciliated epithelium in the lower third of the trachea Considerabl importance is laid on the very frequent occurrence of tracheitis in influenza since destruction and damage to the delicate ciliated epithelium *in this area cause the* disappearance of an effective barrier to invasion of the lungs by inhaled bacteria thus offering an explanation for the great frequency of secondary invasion of the lungs in influenza

The virus may invade the lungs by indirectly gaining access from the congested pharyngeal veins and extravasated blood in the submucosa of *the postnasal region* by traveling in the form of minute infective emboli along the arterial route and may become arrested in the terminal arterioles of the pulmonary artery

CLINICAL VARIETIES The clinical varieties of influenza are separated according to the manifestations of the disease They are (1) simple catarrhal fever (2) pulmonary form (3) gastrointestinal form and (4) nervous form

Simple Catarrhal Fever The usual early symptoms are a feeling of tightness and constriction in the thorax disappearing with the onset of cough and expectoration Rales and rhonchi may be heard in auscultation The simple form tends to be remittent in character the symptoms usually abating in the daytime and increasing in severity toward evening Great prostration attends the disease throughout its course and may last for days or weeks after convalescence

Pulmonary Form The sense of constriction in the thorax dyspnea and stiffness and soreness in the throat are much more intense than in the simple form The

Confirmation of the clinical diagnosis of influenza may be obtained by the demonstration of a rise in the antibody titer of serum collected during convalescence as compared with serum obtained early in the illness and by isolation of virus from nasal or pharyngeal washings It seems that many laboratory workers prefer the agglutination inhibition test This test is based on the fact that influenza viruses agglutinate chicken red blood corpuscles and that this capacity is specifically inhibited by immune serum

Less than a fourfold antibody response formerly was considered within the limits of experimental error The frequent occurrence of twofold increases in titer in the agglutination inhibition test may be considered indicative of influenzal infection However this or any other laboratory test is not available for general diagnostic use in influenza

Differential Diagnosis There are certain types of influenza in which it is often extremely difficult to come to a diagnosis because of their marked resemblance to other infections

Since conjunctivitis laryngitis bronchitis tonsillitis and frontal headache are common to *measles* and influenza a certain diagnosis may not be possible before the fourth day when the fresh febrile accession and the appearance of the rash indicate measles

Owing to the acute onset with shivering and cough influenza may bear a close resemblance to *central pneumonia* from which it can be distinguished by roentgenoscopic examination of the lungs

If the attack of influenza commences with gradual pyrexia with diarrhea rose spots and enlargement of the spleen as happens occasionally in both *typhoid fever* and influenza the diagnosis may have to wait until the temperature begins to fall on the third or fourth day of the illness when the disease is recognized as influenza Leukocytosis is absent in both diseases when uncomplicated

There is often difficulty in the differential diagnosis between the abdominal type of influenza and *acute appendicitis* In acute appendicitis there are increasing numbers of leukocytes whereas in influenza the leukocytes are normal or decreased in number In the absence of a significant leukocyte count the appendicular form of influenza may resemble acute appendicitis so closely as to render a correct diagnosis impossible without a period of observation too long to be safely carried out

Primary Atypical Pneumonia This disease though it occurs as frequently in the older age groups as in the younger has been the subject of written reports chiefly because of its incidence among young adults in schools colleges and army camps

Etiology and Epidemiology A viral etiology for atypical pneumonia has been implied by the exclusive methods practiced by bacteriologists By appropriate methods materials consisting of washings from the noses of selected subjects suffering from primary atypical pneumonia have been transferred to healthy individuals These healthy individuals in due time have manifested respiratory illnesses varying in severity and clinical manifestations corresponding to those of atypical pneumonia

Epidemics of viral pneumonia occur in the fall and winter The disease does not seem to be highly communicable There are numerous instances however of contact infection from patient to nurse and from one member of a family to another These observations suggest that often prolonged and intimate contact with patients is necessary for infection The incubation period is from one to two days No difference has been observed in the susceptibility of the sexes

Pathology Necropsy has revealed patchy hemorrhagic bronchopneumonia with acute bronchitis and bronchiolitis

Symptoms The onset is with a cold or sore throat and cough and chilly feelings but rarely a chill The cough is at first a dry cough A pain through the anterior and midportion of the thorax is almost always present The headache increases in intensity and persists throughout the febrile period The fever tends to be of an irregular type with one or two rises within the 24 hours The temperature peaks

brospinal meningitis, when although the fluid may escape under high pressure it is clear and does not present the characteristic cytologic features of the latter disease

Substernal pain varying from a mild soreness to acute pain is often associated with influenza

In the earlier stages of an epidemic progress is rapid and convalescence is usually established on an average of 5 days without any sequelae Later in an epidemic both the intensity and the virulence of the disease increases convalescence is more protracted as the febrile period lengthens and the bronchitic infections are more pronounced

A high temperature is not a grave symptom during the first 48 hours of influenza Unfavorable symptoms are delirium stupor insomnia persistent vomiting and severe diarrhea

EXAMINATION A bright flushing of the cheeks is common at the commencement of the disease

A light cyanosis is often observed on the cheeks lips ears and neck This coloration is distinctly different from the darker cyanosis commonly seen in pneumonia or bronchitis In those who have capillary bronchitis it may become evident before the development of signs in the lungs

As a peculiarity in influenza *sweating* was observed in early times and the sweating sickness in the sixteenth century may have been a virulent influenza The profuse perspirations occur during the febrile period and may give rise to a curious odor which is said to be similar to that of typhus patients Profuse odoriferous sweating is most often present in very ill patients who have influenza and must be regarded as an unfavorable symptom

A diffusely distributed *erythema* has often been observed *Purpura* may be present as a terminal event Herpes labialis and redness and suffusion about the eyes vary in degree in each case

A relative *slowness of the pulse* is often a common feature while the fever is at its height A sudden rise in the pulse rate indicates a serious complication

The *blood pressure* seems to be at its lowest at about the fourth or fifth day During convalescence the blood pressure rises slowly to its normal but may take a week or two to attain it Much of the feeling of depression and lassitude after influenza is associated with a decreased blood pressure

There are no significant changes in the appearance of the tongue

Vomiting usually ceases during the first 48 hours in the absence of complications or the excessive use of stimulating expectorants or sulfonamides *Diarrhea* often associated with vomiting is common in the first 48 hours

Epistaxis tends to be recurrent rather than obstinate and usually is not a frequent symptom If it is excessive light packing is all that is required If anemia should be found it is not from nosebleed *Hemoptysis* is present if pneumonia is present

No important changes are present in the urine It is scanty in amount highly colored and contains a small amount of albumin

DIAGNOSIS In the sporadic instances of the disease the diagnosis is purely clinical supported by no other evidence The clinical features are considered to be sudden onset of fever muscular aching prostration and various signs of inflammation of the respiratory tract High morbidity rates wide geographic extent and a sharp increase in mortality from pneumonia characterize epidemics many of which during the past century have been recognized on the basis of these criteria *It is* difficult to differentiate the clinical and epidemiologic characteristics of influenza from those of various other forms of respiratory disease

Laboratory methods of diagnosis reveal that the different influenza viruses may produce a rather characteristic form of the disease according to the foregoing criteria as well as numerous mild atypical illnesses and obscure infections

Hyperpnea and cyanosis are present High fever is common in all severe cases Rose spots occasionally appear from the seventh to the thirteenth day Leukocytosis when it occurs is considered to be due to secondary pneumonia from other organisms

A clinical diagnosis is always presumptive The presence of an atypical pneumonia or protracted fever in a patient who has a history of association with sick parrots or canaries justifies a suspicion of psittacosis Once it is suspected that the lung disease is psittacotic a history of association with birds may be obtained The bird is often the canary The virus then can often be isolated by inoculation of citrated blood or sputum into mice

The demonstration of complement fixing antibodies is often employed to establish that a given acute illness is caused by a member of the psittacosis lymphogranuloma group Complement fixing antibodies may appear in the serum from 4 to 8 days after the onset of symptoms Additional serum specimens then are examined from time to time to ascertain a rise in titer within the next 4 or 5 days If there should be rise in the titer it is significant A serum titer of 1 16 or greater is suggestive of psittacosis if the serologic reaction for syphilis is negative and if there is no evidence of infection by the virus of lymphopathia venereum

The mortality rate is about 1 in every 5 of those who have the disease

DISEASES OF THE LUNGS DUE TO HIGHER ANIMAL PARASITES

The selective affinity of parasites for special tissues determines to a large extent the distribution of the lesions

There are several parasitic worms that pass through the lungs during larval development and cause pulmonary disease for instance *Ascaris Strongyloides* hookworm *Trichinella spiralis* and *Echinococcus granulosus* The pulmonary invasion by *Trichinella spiralis* and *Echinococcus granulosus* may leave larvae in the lungs which may cause disease at a later date

Paragonimiasis A parasitic trematode worm or fluke *Paragonimus westermani* selects the lung as the preferred tissue of the host in which to reside and causes the disease known as paragonimiasis

Paragonimus westermani has a wide geographic distribution in the Orient and in South America The adult parasite has been found in mammals in Minnesota Wisconsin Michigan Ohio Mississippi Kentucky South Carolina and northern Canada and in addition the larval forms have been observed in crayfish in Illinois Indiana Iowa Louisiana Missouri Pennsylvania Virginia West Virginia and Ontario Canada Human infestation in North America however is rarely present

Man is infected by eating the raw or improperly cooked flesh of these crustacean hosts

The chief habitat of the adult parasite is the lungs The worms are enclosed in cystic cavities which rupture into the bronchi thereby permitting the ova to reach the sputum Externally the lungs show little change except scattered areas of congestion emphysematous edges and an uneven surface with reddish brown superficial nodules In man these nodules usually number fewer than twenty and do not as a rule project from the surface These nodules are composed of cysts

Paragonimiasis is a slowly progressing chronic condition of insidious onset resembling in many respects tuberculosis Four types have been described (1) general (2) pulmonary (3) abdominal and (4) cerebral

The general type is manifested by fever enlargement of the superficial lymph nodes especially the axillary and inguinal with occasional cutaneous ulceration and in cases of fatal infection muscular rheumatic pains

In the predominating pulmonary type the patients have a chronic cough most pronounced on rising in the morning an abundant blood stained rusty brown purulent tenacious sputum and occasional attacks of hemoptysis usually slight

tend to increase during the first few days of illness but may not reach the maximum until the eighth to the twelfth day Rarely are there dyspnea and cyanosis

Examination On examination there may be a slight impairment of resonance on percussion if the lesion presents at the lung base If the process is central if it presents toward the axillae, or is in the upper pulmonary fields percussion changes may not be found Rales may be heard Physical signs may become detectable only after the acute illness has passed Resolution of the pneumonia is often delayed for weeks This is the so called nontubercular basilar infection of former days

In patients who present no objective physical findings the cold agglutination reaction has been found to be of some value in the diagnosis of this disease The cold agglutination reaction is based on the presence of agglutinins which cause clumping of homologous or group O red cells at low temperatures The test is not infallible since not all patients yield a positive reaction Furthermore cold agglutinins have been reported in paroxysmal hemoglobinuria, trypanosomiasis pernicious anemia leukemia lymphoblastoma cirrhosis venous thrombosis and gangrene among others

The leukocyte count is normal or there may be a leukopenia Leukocytosis if present may appear after several days The sputum does not contain predominating bacteria Cold agglutinins appear during the second week in a majority of cases of primary atypical pneumonia and therefore are of no great diagnostic assistance

Diagnosis On general examination there may not be any pulmonary signs Roentgenologic examination may present the first evidence of pulmonary involvement The lesion seems to begin at the hilus of the lung and spreads outward However in some instances roentgenologic examination does not reveal changes in the lungs during the active course of the disease The disease is revealed after the symptoms have subsided

In young adults the prognosis is good In older patients whom the disease affects like bronchopneumonia the outcome is more likely to be fatal

There is no effective treatment Chemotherapy and antibiotic therapy are worthless or harmful

Psittacosis and Ornithosis (Psittacosis Lymphopathia Viruses) The infective agents of psittacosis of birds and man lymphopathia venereum of man and murine and feline pneumonitis seem to be disease agents intermediate between the rickettsias and viruses These belong to the so called chlamydozoa and are gram negative organisms

Psittacosis Synonyms for psittacosis are ornithosis parrot fever psittacose and Papageienkrankheit The avian psittacosis is caused by an elementary body virus which develops in reticuloendothelial cells It is communicable to man

Psittacosis is an acute and sometimes protracted febrile illness with pulmonary lesions which is transmitted from sick parrots to man A viral disease with similar pathologic manifestations is found in wild birds and tamed canaries as well as in pigeons and chickens

The disease is severe has a high mortality rate and affects all ages and both sexes The incubation period is usually about 10 days in exceptional cases as short as 4 or as long as 16 days

The pathologic changes observed are those of septicemia with localizing pulmonary lesions

Headache is a constant symptom and epistaxis often occurs There are generalized aches and pains sore throat and herpes labialis Chilliness chills and rigors are frequent The cough is severe and paroxysmal The sputum is scanty mucoid sometimes purulent occasionally blood stained but infrequently rust colored Abdominal pain nausea and vomiting are common

On examination after the fifth day of illness a few crepitant rales are present in the lungs Later signs indicate patchy regions of consolidation in one or both lungs

the tissues become cyanotic and peripheral edema appears All of these changes are indicative of chronic passive congestion

It may be inferred from the causes which originate chronic passive congestion of the lungs that essentially this process is one of chronic edema An acute pulmonary edema may arise during the course of such a chronic passive congestion and the latter constitutes one of the common causes of acute pulmonary edema

Acute Pulmonary Edema This is a serious condition which may be the immediate cause of death in cardiac and circulatory failures such as acute and chronic failure of the left ventricle acute coronary thrombosis angioneurotic edema and the excessive intravenous administration of fluids Acute pulmonary edema may arise from extensive acute pulmonary disease such as pneumonia extensive malignant metastasis and inflammation produced by the inhalation of irritant gases

The symptoms are the manifestations of lungs filled with fluid with all of the anxieties and apprehensions of an individual drowning Add to this the agony of pain such as is present in the case of cardiac infarction and there is truly the combined picture of horror and death

On examination there is profound cyanosis of the face and extremities The superficial veins of the face neck and upper extremities are distended The extremities are often dark and cold Pressure applied to the skin of such an extremity leaves a blanched area which remains white for some time

The vibrations from the audible rhonchi and rales are palpable On percussion the note is flat over the posterior portions of the lower lobes There are so many noises present in the lungs that auscultation is not helpful diagnostically

The diagnosis is usually obvious from inspection The roentgenoscopic examination often gives some clue to the physical state of the heart lungs pleura and mediastinum

Pulmonary Embolism Embolism is the sudden obstruction of the lumen of a blood vessel by a blood clot air fat or any other substance which has been brought to its place by the blood current

The commonest embolus reaching the larger pulmonary arteries is composed of clotted blood Such blood clots may arise in the heart affected by valvular heart disease or in the larger veins usually those of the legs In the leg veins there are two conditions prone to create emboli These conditions are phlebothrombosis and phlebitis In phlebothrombosis there is no evidence of antecedent disease or injury of the vein Thrombosis is first The phlebitis develops as a reaction to the thrombosis

Embolism usually in the lungs is the most serious complication of postoperative venous thrombosis It is often the result of phlebothrombosis and occurs in many cases in which there is no clinical evidence of thrombophlebitis Therefore it is assumed that thrombosis has occurred in some one of the peripheral veins and that either the entire thrombus became the embolus or the part of it which remained at the original site failed to produce local symptoms

Surgical exploration for inoperable carcinoma associated with metastasis is accompanied with high incidence of postoperative pulmonary embolism Usually the magnitude of the surgical procedure is directly related to the frequency of embolism which is twice as common after repair of bilateral hernia as after operations for unilateral hernia

Thrombophlebitis predisposes to pulmonary embolism but there are reasons for believing that the clinical manifestations of peripheral thrombophlebitis are associated with fixation of the thrombus by inflammation

When pulmonary embolism occurs after the clinical manifestations of thrombophlebitis have appeared the emboli are likely to be small and nonfatal It is commoner to observe phenomena of pulmonary embolism a few days before clinical thrombophlebitis than after the symptoms of thrombophlebitis have appeared

though at times severe The physical signs may suggest lobular pneumonia or tuberculosis

A definite diagnosis is established by finding the ova in the sputum the feces or in the cutaneous or subcutaneous tissues of a patient who has pulmonary disease

In light infections prognosis is good although spontaneous cure may not occur *In heavy invasions* superimposed tuberculosis secondary pyogenic infections or cerebral involvement the prognosis is unfavorable

Echinococcus Cysts of the Lungs Echinococcus cysts of the lungs are caused by *Echinococcus granulosus* of the superfamily Taenioidae

The *symptoms* if any are present are cough hemoptysis dyspnea and pain The cough is dry and hacking and may be accompanied by slight mucoid expectoration Hemoptysis in small quantities may occur Mild thoracic discomforts but rarely pain unless the pleura is reached may develop as may a pleural rub also In pleural involvement there may be a low grade fever The cyst may give pressure symptoms which are followed by rupture into a bronchus or it may penetrate the pericardium a chamber of the heart the vena cava or the thoracic wall

The rupture of a cyst into the bronchial tree originates sudden distress pain and the expectoration of quantities of thin blood stained fluid If the patient survives the rupture the diagnosis of hydatid cyst becomes evident as fragments of cyst are coughed up After rupture urticaria is common Sudden death has followed the impaction of fragments of the cyst in the glottis Profuse hemoptysis may occur at the time of rupture or later and may prove fatal

On examination over the cyst fremitus is diminished or absent Percussion dullness may be elicited There is often displacement of mediastinal structures Auscultation may reveal changes in respiratory sounds because of the space occupying cyst Crackling or musical rales are common Occasionally percussion and auscultation may be altered by the effects of a bronchial communication Roentgenologic examination reveals the cysts

The diagnosis is made by identification of the parasite or the cysts The intradermal and the complement fixation tests may be helpful The clinical manifestations and the roentgenologic evidence are often diagnostic Puncturing and aspiration of the cysts are contraindicated because of the danger of rupture and anaphylactic phenomena If the situation of the cyst is favorable for surgical approach operation should be considered

DISEASES OF THE LUNGS SECONDARY TO CIRCULATORY DISTURBANCES

Acute and Chronic Pulmonary Congestion Acute and chronic pulmonary congestion occurs in certain primary and secondary diseases of the lungs The primary diseases of the lungs which are characterized by acute congestion are the acute infections and the acute injuries from physical and chemical agents Acute congestion from primary disease of the lungs may be localized as in the pneumococcal pneumonias or generalized when provoked by the inhalation of an irritating gas

Chronic congestion of the lungs occurs less frequently from primary pulmonary disease than it does from disease in which the lungs are secondarily congested as in heart failure from disease of the mitral valves Chronic passive congestion of the lungs as it occurs in mitral heart disease renders the lungs less elastic and more rigid than normal The lining of the bronchial tubes is malnourished and desquamates and thus permits fluid and erythrocytes to exude from the capillaries into the air passages to be expectorated Cough and increased expectoration may be the only complaint that a patient has for seeking help However before long the patient has dyspnea and the sputum becomes reddish brown (rusty) in color the peripheral veins are dilated the capillaries are widened and pulsate the skin of the lips and

the tissues become cyanotic and peripheral edema appears All of these changes are indicative of chronic passive congestion

It may be inferred from the causes which originate chronic passive congestion of the lungs that essentially this process is one of chronic edema An acute pulmonary edema may arise during the course of such a chronic passive congestion and the latter constitutes one of the common causes of acute pulmonary edema

Acute Pulmonary Edema This is a serious condition which may be the immediate cause of death in cardiac and circulatory failures such as acute and chronic failure of the left ventricle acute coronary thrombosis angioneurotic edema and the excessive intravenous administration of fluids Acute pulmonary edema may arise from extensive acute pulmonary disease such as pneumonia extensive malignant metastasis and inflammation produced by the inhalation of irritant gases

The symptoms are the manifestations of lungs filled with fluid with all of the anxieties and apprehensions of an individual drowning Add to this the agony of pain such as is present in the case of cardiac infarction and there is truly the combined picture of horror and death

On examination there is profound cyanosis of the face and extremities The superficial veins of the face neck and upper extremities are distended The extremities are often dark and cold Pressure applied to the skin of such an extremity leaves a blanched area which remains white for some time

The vibrations from the audible rhonchi and rales are palpable On percussion the note is flat over the posterior portions of the lower lobes There are so many noises present in the lungs that auscultation is not helpful diagnostically

The diagnosis is usually obvious from inspection The roentgenoscopic examination often gives some clue to the physical state of the heart lungs pleura and mediastinum

Pulmonary Embolism Embolism is the sudden obstruction of the lumen of a blood vessel by a blood clot air fat or any other substance which has been brought to its place by the blood current

The commonest embolus reaching the larger pulmonary arteries is composed of clotted blood Such blood clots may arise in the heart affected by valvular heart disease or in the larger veins usually those of the legs In the leg veins there are two conditions prone to create emboli These conditions are phlebothrombosis and phlebitis In phlebothrombosis there is no evidence of antecedent disease or injury of the vein Thrombosis is first The phlebitis develops as a reaction to the thrombosis

Embolism usually in the lungs is the most serious complication of postoperative venous thrombosis It is often the result of phlebothrombosis and occurs in many cases in which there is no clinical evidence of thrombophlebitis Therefore it is assumed that thrombosis has occurred in some one of the peripheral veins and that either the entire thrombus became the embolus or the part of it which remained at the original site failed to produce local symptoms

Surgical exploration for inoperable carcinoma associated with metastasis is accompanied with high incidence of postoperative pulmonary embolism Usually the magnitude of the surgical procedure is directly related to the frequency of embolism which is twice as common after repair of bilateral hernia as after operations for unilateral hernia

Thrombophlebitis predisposes to pulmonary embolism but there are reasons for believing that the clinical manifestations of peripheral thrombophlebitis are associated with fixation of the thrombus by inflammation

When pulmonary embolism occurs after the clinical manifestations of thrombophlebitis have appeared the emboli are likely to be small and nonfatal It is commoner to observe phenomena of pulmonary embolism a few days before clinical thrombophlebitis than after the symptoms of thrombophlebitis have appeared

Allen Barker and Hines have stated that when a patient has a pulmonary embolism after operation and survives there is a 43 8 per cent chance of another thrombotic episode a 30 5 per cent chance of another embolism and an 18 3 per cent chance of a fatal embolism in the same postoperative convalescence The incidence is less in cases of thrombophlebitis than in cases of phlebothrombosis but is still definitely higher than in all postoperative cases as a group

Pulmonary embolism affects men much more frequently than women, despite the preponderance of operations on the lower part of the abdomen among women

SYMPTOMS The symptoms depend on the amount of lung tissue which is incapacitated by the occluded artery The occlusion of a small pulmonary artery may occur without noticeable symptoms The occlusion of a relatively large pulmonary artery produces cor pulmonale and shock The occlusion of a large or several small pulmonary arteries may be immediately fatal

On occasion the patient who has a pulmonary embolism will have a sudden feeling of apprehension anxiety palpitation and dyspnea These symptoms are often so severe that the patient is fearful of moving The duration of the attack is often from 15 to 30 minutes Several hours or a day or two elapse before pleuritic pain or hemoptysis occurs

Pleuritic pain which occurs in about one week to 10 days after an operation is often caused by pulmonary embolism A distinctive feature of pulmonary embolism often is dyspnea out of proportion to restriction of respiratory movements and associated pain The pain occurs as the result of the embolus reaching a peripheral pulmonary artery and thus involving the pleura Despite pain a small embolus gives no general symptoms and the patient maintains a sense of well being

Hemoptysis occurring after operation should be regarded as evidence of pulmonary embolism The patient coughs up small blood clots or expectorates a thick bloody sputum As time passes the coughed up blood becomes darker and decreases in amount

Small pulmonary emboli with minimal or no symptoms often occur This occurrence indicates that the patient is susceptible to thrombosis and resultant embolism Once this process is initiated it will recur often and there is danger of subsequent massive pulmonary embolism

In *massive pulmonary embolism* there is an initial shock The initial shock of a large pulmonary embolus may be survived and if so a prolonged febrile illness may follow The illness results from a large pulmonary infarct which may or may not break down to produce pulmonary abscess This condition resembles pneumonia and is difficult to differentiate from pneumonia

EXAMINATION The findings on physical examination after pulmonary embolism are exceedingly variable and often inconclusive Rales percussion dullness and pleuritic friction sounds are irregularly present in the beginning As time passes the physical signs of a localized pneumonia may appear The presence of unilateral pleural effusion after a surgical operation is highly suggestive that pulmonary embolism has occurred

Examination soon after a large pulmonary embolus has occurred reveals evidence of shock In smaller but definitely manifested emboli there are present dyspnea perhaps orthopnea and always varying degrees of apprehension If a large area of the lung has been embolized there is an increased pulsation in the second and third interspaces to the left of the sternum and a loud systolic murmur with accentuation of the second pulmonic sound Gallop rhythm is present occasionally The veins of the neck are dilated and pulsating There is an initial fall of temperature and then a rise to 101 or 103 F (38 3 or 39 4 C) as the evident degree of shock abates There may be scattered or numerous moist rales in the lungs and the breath sounds may be distant or suppressed over the affected portion of the

lung Signs of the local accumulation of fluid in the pleural space may develop after a few days Rather severe and persistent hemoptysis often is evident

There is no characteristic roentgenographic evidence of a large pulmonary infarct Large infarcts frequently produce elevation of the diaphragm on the affected side indefinite thickening of the pleural shadow in the costophrenic angle and occasionally the appearance of a small amount of pleural fluid

Electrocardiographic changes which are indicative of acute cor pulmonale occurring with severe pulmonary embolism may be present The changes in the electrocardiogram may develop at any time during the first 48 hours after the embolism occurs and they may be transient but when they are found they are significant (Barnes)

A large pulmonary embolus is one of the causes of sudden death Some patients may live for a few minutes after the episode or more rarely a few hours may elapse before death occurs

DIAGNOSIS The occurrence of sudden pleural pain from 6 to 21 days after an operation obstetric delivery or severe injury should be regarded with great suspicion since other conditions causing pleurisy are rare in those periods Sudden death occurring in the same group of patients during the same interval should be considered as most likely the result of a large pulmonary embolus particularly if the clinical course has otherwise been normal If pleural pain hemoptysis and shock as produced by a relatively large pulmonary embolus occur in a patient who has acute or recent thrombophlebitis the diagnosis of embolism can almost certainly be made but as has been stated most embolisms occur before thrombophlebitis is clinically evident

The one condition difficult at times to differentiate is an acute coronary thrombosis The electrocardiogram is of great value in distinguishing acute coronary thrombosis from pulmonary embolism since the patterns are different even if the coronary thrombus is in the posterior descending branch In coronary thrombosis frequently the pain is more severe it is not pleural it often extends to the arms or shoulders cyanosis is infrequent and signs of a localized pulmonary lesion are absent

The electrocardiographic changes in infarction of the posterior ventricular wall and those present in pulmonary embolism are somewhat similar The deep S and the inversion of the T waves present after pulmonary embolism do not occur in infarctions of the posterior ventricular wall

The signs and symptoms of small pulmonary infarcts usually disappear within a week with the possible exception of some residual elevation of the diaphragm on the affected side

Air Embolism Air embolism may occur in two forms (1) pulmonary venous air embolism and (2) arterial air embolism

In the pulmonary form air enters one of the systemic veins and is carried to the right side of the heart and pulmonary circulation and depends for its effects on mechanical obstruction of the outflow channel of the right ventricle

In arterial embolism the air gains entrance to the pulmonary venous channels and is propelled from the left ventricle to those systemic arteries which supply the superiorly located portions of the body Since a small amount of air can effectively block a medium sized artery serious consequences may result when a few milliliters of air enter the pulmonary vein and are carried into either the coronary or cerebral circulation or both

In the pulmonary form the presence of air in the right ventricle produces a loud churning sound often readily heard without stethoscopic aid which is known as the millwheel murmur This murmur appears almost immediately after air has entered the venous circulation

expectoration are absent except when conditions such as bronchial infections or asthma are present Cyanosis and compensatory polycythemia may appear

EXAMINATION Inspection in severe emphysema reveals the thorax permanently in the position of deep inspiration with slight movements of the thoracic wall the phrenic wave sign is absent The lungs have lost their elastic recoil so that the intercostal and abdominal muscles are used for breathing Owing to the voluminous lungs the diaphragm is stretched and its function is impaired Therefore the neck and intercostal muscles may actively support inspiration The action of the latter muscles pulls the thoracic wall upward and forward The expanded lungs extend into the pleural sinuses and between the heart and thoracic wall resulting in increase of the anteroposterior diameter of the thorax and dorsal kyphosis

If the emphysema is severe, cyanosis may be observed and the veins of the neck are distended In some cases there is clubbing of the fingers and toes

On palpation decreased expansion of the thoracic wall and diminished tactile fremitus are evident The percussion note is hyperresonant and the lower margins of the lungs extend below their normal limits The area of cardiac dullness is diminished and the liver is in a low position On auscultation the expiratory phase of respiration is prolonged and low pitched Whispered voice sounds are diminished Rales are elicited only when other conditions such as asthma and bronchitis are present

DIAGNOSIS The appearance of the patient in combination with the physical and roentgenologic findings is diagnostic

Many persons endure emphysema over long periods of time

Senile Emphysema Elderly persons who have kyphosis of the thoracic vertebrae and some who do not have deformities of the spinal column may have a general narrowing of the thorax and the intercostal spaces and a decrease in the respiratory movements of the thoracic wall without symptoms

On examination tactile fremitus is diminished the percussion note is hyperresonant and auscultation reveals diminished breath sounds Often only the expiratory phase of respiration is heard

The diagnosis is made during routine physical examinations

Traumatic or Interstitial Emphysema Rupture of alveolar walls may result from needle puncture crushing or tearing injuries of the chest violent coughing and from the use of a pulmotor Interstitial emphysema may develop in those who are enjoying health without evidence of disease (spontaneous interstitial emphysema)

The escaped air from the lung may spread through the interstitial tissue and through the hilus and thence into the mediastinum (*mediastinal emphysema*) thence it may spread into the tissues of the neck and occasionally gives rise to generalized subcutaneous emphysema

SYMPTOMS When the amount of air is small there are no symptoms When a large amount of air escapes some of it may dissect along the connective tissue bands surrounding the blood vessels and form blebs on the pleura When a bleb ruptures spontaneous pneumothorax occurs

In some instances the first symptom is severe pain beneath the sternum or in the side of the thorax The pain may extend to the left side of the neck and down the left arm The pain of spontaneous interstitial emphysema disappears soon after the attack

EXAMINATION The late Louis Hamman described as being characteristic of this condition a peculiar crackling crunching sound heard over the sternum and precordium which is synchronous with the heart beat This sound may be heard with the patient in any position but it is more likely to be elicited when the patient lies on the left side or leans forward These sounds convey the impression of air being churned or squeezed in the tissues Often they are so loud that they can be heard by the patient Interstitial emphysema causes a hyperresonant note to percussion over

the usual region of cardiac dullness In some cases signs of spontaneous pneumothorax occur

DIAGNOSIS The history and the examination by physical means are diagnostic The results of roentgenologic examination are negative unless pneumothorax is present

Symptoms soon disappear

TUMORS OF THE LUNG

Tumors of the lung may be arranged in certain groups based partly on the manner of origin and partly on the tissue from which the tumor originates (1) Bronchial malformations may be productive of benign cystic and papillary structures The congenital cystic malformations are of two types (a) overgrowth of misplaced fetal portions of the bronchial tree and (b) overgrowth of areas of fetal bronchiectasis resulting from inflammation and leading to occlusion of a bronchus and retention of mucus (2) Mixed tumors also of fetal origin arise from fetal pulmonary tissues which fail to develop into normal pulmonary structures These tumors may be designated as chondromas or sarcomas When the epithelial elements predominate they are termed adenoma alveolar carcinoma or adenocarcinoma Hamartoma of the lung may contain cartilage and is thus related to the chondromas However not all hamartomas contain cartilage (3) The myxomas and fibromas of the periphery of the lung probably arise from the pleura (4) The solid diffuse carcinomas which arise as a sequel or incident to inflammations such as a chronic tuberculous pneumonia are difficult to separate from bronchial carcinoma

Regardless of the origin of lung tumors which may be classified as being benign tumors they are comparatively rare The so-called bronchial adenoma and the chondroma or hamartoma are the commonest benign tumors of the lung

The detection of these lung tumors is accomplished by roentgenologic examination The diagnosis awaits histologic study The prognosis of course depends on the degree of anaplasia of the cells

Metastatic Malignant Disease of the Lung Metastatic malignant lesions of the lungs are many times more frequent than primary ones

Epithelial Malignant Lesions Carcinomas hypernephromas and chorionepitheliomas almost always give secondary pulmonary involvement Carcinomas of the breast frequently metastasize to the lungs Malignant lesions of the thyroid gland air passages alimentary tract and its associated glands the urinary organs genitalia and extremities often aid in the destruction of the patient through metastasis to the lungs

Pulmonary metastasis occurs by (1) direct extension (2) blood stream implantation and (3) lymphatic spread The metastatic lesions usually involve the lower portions of the lungs They appear as (1) single or multiple nodular growths in one or both lungs and (2) infiltrative or diffuse growths in both lungs A lung is more completely filled with secondary malignant disease in instances of extension from cancer of the breast than in extension from cancer elsewhere The affected lung usually corresponds to the breast involved by carcinoma The pleura is often extensively involved as shown by a hemorrhagic or serofibrinous effusion

SYMPTOMS A few who have metastatic pulmonary carcinoma may have cough vague thoracic discomforts and at times pleuritic pain Dyspnea may be a distressing symptom and hemoptysis is common However on roentgenologic examination the lungs are frequently found to be involved without there having been any symptoms or findings on examination of the thorax by physical means

EXAMINATION In early periods of metastatic pulmonary tumors there may be observed a suppression of the breath sounds especially pronounced during inspiration and sometimes accompanied by fine crackling rales over a limited area of the thorax As the tumor or tumors grow decreased expansion ensues The percussion

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note then becomes flat and bronchovesicular or bronchial breathing and sibilant and sonorous rales are present Signs of pleural effusion may be found early or at least during the course of the disease These signs are usually manifest over the lower half of the lungs particularly

DIAGNOSIS Roentgenologic examination is usually diagnostically conclusive It is well to try to confirm the diagnosis if possible by *repeated cytologic examinations* of the sputum Bronchoscopy is of little or of no diagnostic value

Sarcomas Metastatic involvement of the lungs by sarcomatous tumors is proportionately commoner than by carcinomatous tumors Sarcomas arising in almost any part of the body metastasize and secondarily involve the lungs

SYMPTOMS In metastatic pulmonary sarcoma as in other pulmonary malignant diseases both primary and secondary cough pain in the thorax dyspnea and hemoptysis are the usual symptoms

EXAMINATION When metastatic nodules have attained considerable size in the lungs dullness or flatness to percussion is elicited Sibilant and sonorous or musical rales are heard and the breath sounds are decreased and changed to a bronchial tone *Massive growths obliterate all resonance breath sounds and vocal* fremitus or transmission of voice sounds over an entire lung field or over most of the thorax even in the absence of sufficient fluid to produce such signs In some cases bulging or retraction of the thorax or the intercostal spaces may be visible

DIAGNOSIS The presence of metastatic pulmonary malignant disease in a patient known to have a sarcomatous tumor in any part of the body is assumed to be pulmonary sarcoma unless proved to be otherwise The physical signs and the roentgenoscopic findings are diagnostic of a malignant tumor of the lung only not of the type of tumor

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10

DISORDERS OF THE HEART, BLOOD PRESSURE, PULMONARY ARTERIES AND THORACIC AORTA

THE HEART

The average adult heart measures about 5 inches (12 7 cm) in length 3 inches (7 6 cm) in width and 2½ inches (6 4 cm) in thickness The average weight of the healthy adult heart of a man is about 9 ounces (280 gm) and of a woman 8 ounces (250 gm) These weights may be doubled in instances of cardiac enlargement

Outlines of the Heart The base or upper margin of the heart from which the great vessels go off lies in the second intercostal space or at the upper margin of the third rib behind the sternum It is here that the superior vena cava ends and the aorta begins The base of the heart extends from about ½ inch (1 3 cm) to the right of the sternum to about 1 inch (2 5 cm) to the left of the sternum

The *right margin* of the heart formed by the right atrium lies from about 1½ to 1¾ inches (3 8 to 4 5 cm) to the right of the median line at the upper border of the third costal cartilage and extends in an outwardly curved line to the junction of the seventh rib and the sternum In the fourth interspace it may reach 1 inch (2 5 cm) beyond the right edge of the sternum

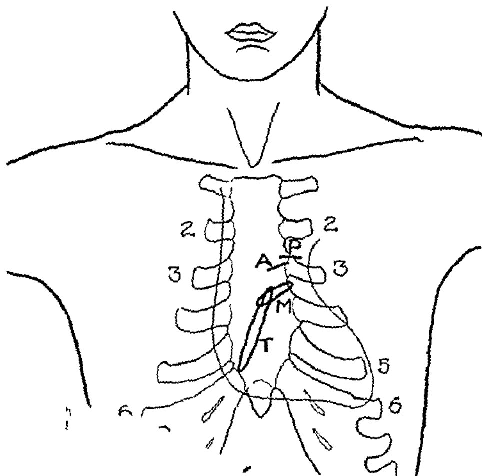
The *left margin* of the heart passes somewhat obliquely downward from 1 inch (2 5 cm) to the left of the sternum at the upper border of the third costal cartilage and to the left to the region of the apex

The curved sternocostal surface of the heart which proceeds upward and forward and comes into direct contact with the anterior wall of the thorax is formed chiefly by the right atrium and the right ventricle it lies just behind the sternum and behind the anterior extremities of the third fourth fifth and sixth ribs being in part overlapped by the margins of the lungs

The *lower margin* of the heart passes from the seventh right chondrosternal junction across the sternoxiphoid joint outward in the fifth interspace to the apex beat which is from 1⅗ to 2 inches (4 1 to 5 1 cm) below and to the inner side of the nipple and about 3½ inches (8 9 cm) to the left of the median line This marks the extreme left limit of the heart In children the apex is higher in the fourth interspace In old people it is lower

The diaphragmatic surface of the heart posterior and inferior is almost flat where it is in contact with the diaphragm The left atrium lies at the back of the heart and is directed toward the esophagus and spinal column the left ventricle lies behind and below only a small portion of it projecting forward to form the apex of the heart The auricle of the left atrium is in contact with the anterior wall of the thorax close to the pulmonary artery The orifice of the pulmonary artery is situated at the sternal end of the third left intercostal space the orifice of the aorta lies just below the middle of the left half of the sternum at the same level The center of the mitral orifice lies behind the third left intercostal space close to the sternum and the center of the tricuspid orifice lies behind the right half of the sternum at the level of the sternal end of the fourth intercostal space (Fig 10 1)

The *atrioventricular (auriculoventricular) groove or line of junction between* the atria (auricles) and ventricles runs from the sixth right chondrosternal junction upward and to the left to the third left chondrosternal junction. The atria lie above and to the right of this line and the ventricles below and to the left.



of examination (1) the inspection and palpation of the apex beat of the heart (2) the percussion of the cardiac dullness and (3) the delimitation of the margins of the heart by means of roentgenologic examination

Position of the Apex Beat of the Heart In the healthy and not too fat man an elevation of the thoracic wall at regular intervals at the times of the systoles of the left ventricle can be seen and felt in the left fifth intercostal space between the mammillary and the parasternal lines When the heart is enlarged the apex beat changes its position it may occupy the sixth seventh or eighth interspace instead of the fifth and may be as far as 2 or 3 inches (5 1 to 7 6 cm) to the left of the nipple line

The site of the apex beat is the most lateral and the lowest part of the area of pulsation The apex beat thus recorded according to H L Smith does actually correspond to the position of the heart s apex Thus the point of maximal impulse (p m i) is almost always at the site of the apex beat

The position of the apex beat with the patient standing is practically the same as in the recumbent position On deep inspiration the apex beat may descend and lie behind the sixth rib or in the sixth intercostal space on deep expiration in the young and healthy person it may rise as high as the fourth intercostal space and become more diffuse The position changes when the patient lies on his left side it may then reach the mammillary line or often a point to the left of it After rapid emaciation this mobility may be greater when the patient lies on his right side the apex is very little displaced to the right rarely passing to the right of the left parasternal line

The position of the apex beat varies somewhat with the shape of the thorax in the different somatic types and as the results of thoracic deformities Anything that lowers the diaphragm lowers the apex beat also anything that raises the level of the diaphragm (meteorism ascites pregnancy abdominal tumors) will raise the position of the apex beat Fluid or air in one pleural cavity will displace the apex beat toward the opposite side retraction of a lung may have an opposite effect Enlargement of the left ventricle displaces the apex downward and to the left enlargement of the right ventricle displaces the apex to the left but not downward A pericardial effusion may displace the apex beat upward to the third intercostal space

Too much stress should not be laid on slight abnormalities in the position of the apex beat Its determination however is the first step in the attempt to ascertain the position and size of the heart inasmuch as the position of the apex indicates the situation of the left margin of the heart

Precordial boss and voussure are the names given to a unilateral bulging occupying the precordial area in children This bulging is due to enlargement of the heart or to pericardial effusion The enlargement is oval its long diameter being vertical and extending from the third to the sixth rib near the sternum It can arise only in early life while the ribs are still flexible

Percussion of Areas of Cardiac Dullness By percussion on the anterior surface of the thorax the area of cardiac dullness often can be outlined particularly in young and healthy persons Two areas of cardiac dullness may be delineated These areas are (1) that of the superficial small cardiac dullness and (2) that of the deep or relative cardiac dullness The area of small dullness corresponds to the part of the sternocostal surface of the heart not covered by the lungs while the area of relative dullness corresponds to the whole heart in orthogonal projection The area of superficial dullness is outlined by feeble percussion along the margins of the right and left lungs where they overlap the heart Inside the boundary of this area the characteristic resonant pulmonary note is absent The larger area of relative dullness is outlined by percussing from the lungs toward the heart and marking the points at which the percussion note becomes less resonant or relatively

dull Though the heart is covered by the lungs at its periphery the change in the percussion sound elicited after practice and experience permits the determination of approximate outlines of the heart with reasonable accuracy The determination of the superficial dullness is much easier than determination of the deep dullness, for the latter a degree of skill obtainable only by considerable practice is required

In obesity and in emphysema the determination of the relative dullness is difficult if not impossible The area obtained is much smaller than that corresponding to the true dimensions of the heart When the dullness passes to the right as far as the right parasternal line or farther and upward as high as the second rib while the left margin of the heart is as far as or farther to the left than normal it is certain that the heart is enlarged

Dilatation principally of the left ventricle causes an expansion of the area of dullness toward the left only Dilatation of the right ventricle displaces the right margin of the area of relative dullness to the right When both relative and absolute dullness are increased to the right of the sternal margin there is usually either a dilatation of the right atrium or a pericardial exudate

The apparent area of cardiac dullness as outlined by percussion may not be cardiac dullness at all but may result from the presence of tumors in the neighborhood of pleural effusions or of infiltrations of the lungs Similarly a dislocation of a heart of normal size by a pleural effusion by a tumor or by a pneumothorax may simulate an abnormal position of one cardiac boundary due to enlargement

Diminution of the Area of Dullness Seldom does this indicate diminution in size of the heart often it is due to emphysema

The percussion of the heart is a subjective method of measuring the heart which has much value though the error is often considerable A high degree of accuracy in percussion is impossible for many reasons There are no sharp margins of cardiac dullness to be obtained by percussion in the various intercostal spaces No outlines of the heart by percussion are as important diagnostically as the finding of a well defined point of maximal impulse The value then of percussion of the heart lies mainly in enabling confirmation of the suspected left limits of the heart when the impulse is ill defined This feat is often of enough value to reward for the time spent in the practice of percussion and much time is required

Normal and Abnormal Sounds Over the Heart and Vessels Obtained on Auscultation In health and in disease oscillations arise in the heart and blood vessels that may cause audible sounds These can be recognized by listening with the naked ear or with the aid of the stethoscope

THE HEART SOUNDS If the ear or a stethoscope is placed over the thorax in the cardiac region of a healthy person pairs of sounds are heard recurring at regular intervals Each pair of sounds consists of a first and a second sound separated from each other by a brief interval Each pair of sounds in turn is separated from the next pair by a longer interval The duration and accentuation of the single sounds at the apex of the heart are found to be somewhat different from the duration and accentuation at the base thus at the base and over the lower part of the sternum the first sound is longer and more accentuated phonetically indicated as *lubb dupp* whereas at the apex the second sound is more accentuated the rhythm becoming that of *lubb dupp* The first sound of each pair is synchronous with the apex beat of the heart The second sound occurs at the very beginning of ventricular diastole The long pause separating consecutive pairs is diastolic in time

Origin of the Normal Heart Sounds The origin of the *first sound* has been shown by H L Smith Essex and Baldes to be chiefly due to closure of the mitral and tricuspid valv[illegible] is an element of intracardial tension tone due to oscillations [illegible]ions of the walls of the ventricles it is less important in au[illegible]

The *second sound* is coincident with and due to the closure and tension of the semilunar valves of the aorta and pulmonary artery at the beginning of diastole

In children and young adults occasionally the trained ear may hear a *third sound* audible just after the second sound It is said to be best heard at or near the apex when the patient is somewhat turned to the left side

Auscultation Sites The sites selected for auscultation are those at which the vibration of the particular valves in question is best heard These auscultation sites for the pulmonary and tricuspid valves correspond to anatomic projections of the valves The auscultation sites for the aortic and mitral valves do not correspond to the surface overlying the anatomic sites of these valves

The four principal auscultation sites are (1) at the apex of the heart for the mitral valves (mitral area) (2) at the second left intercostal space close to the sternum for the pulmonary semilunar valves (pulmonary area) (3) at the second right intercostal space close to the sternum for the aortic valves (aortic area) and (4) at the junction of the fifth rib with the sternum on both the right and left sides for the tricuspid valve (tricuspid area)

Rhythm and Accentuation At each auscultation site both sounds of the heart are audible and experience and training teach the art of distinguishing at each site the sound of autochthonous origin from the propagated sound Thus over the pulmonary and aortic sites the second sounds are autochthonous and the first sounds are propagated

Identification of the Heart Sounds Over a healthy heart the distinguishing of the first sound from the second sound is readily accomplished by the accentuation and by the length of the pause following each sound In disease however the heart beats may become very irregular or the rate greatly accelerated Both the long pause and the first sound become relatively shortened and the heart sounds are like those of a fetus so that the ear may not be able to distinguish the first from the second sound In such a case the examiner may help himself by palpating (simultaneously with auscultation) the apex beat or the carotid pulse

The loudness of the sounds of the heart may be less or greater than normal

Enfeeblement of the First Sound The first sound is enfeebled when something exists that hinders the transmission of the sound such as obesity pulmonary emphysema large pleural or pericardial effusion defective contraction of the ventricles or alterations of the atrioventricular valves A feeble contraction of the ventricle may occur in any of the conditions that give rise to myocardial insufficiency When vegetations appear as in acute endocarditis on the borders of the valves the first sound may be diminished or may even disappear

Accentuation of the First Sound The first sound may be exaggerated in intensity when the transmission of the sound is facilitated as in emaciated patients or when there are gas-containing cavities near the heart (pneumopericardium dilated stomach) Abrupt ventricular contraction such as often occurs in emotional excitement or on physical exercise augments the first sound and certain alterations in the atrioventricular valves such as thickening or induration may have the same effect The latter may explain the loud first sound sometimes heard in chronic endocarditis for example mitral insufficiency and in arteriosclerosis of a mitral cusp

A marked accentuation of the first sound at the apex often indicates the existence of mitral stenosis The abrupt first sound heard in this disease is believed to be due to the more powerful vibrations set up by the sudden tension of the stiffened valve cusps

Enfeeblement of the Second Sound The second sound may be feebler than normal owing to faulty transmission to the stethoscope as in obesity pulmonary emphysema and pericardial effusion and owing to lowering of tension in the aorta or pulmonary artery as in myocardial insufficiency anemia hypotension and in general debility

The lesion of mitral insufficiency tends to enfeeble the first sound When in the course of a mitral stenosis a mitral insufficiency develops the first sound may become somewhat feebler

Accentuation of the Second Sound The second sound may be exaggerated (accentuated) whenever the arterial tension is increased Thus marked accentuation of the aortic second sound usually indicates hypertension in the aorta

When in addition to accentuation of the aortic second sound there is alteration of the quality or timbre there often are changes in the semilunar valves themselves in which case the sound is loud and has a certain tympanitic or ringing quality

When the accentuation of the second sound is maximal to the right of the sternum it is certainly aortic in origin but when it is maximal to the left of the sternum it is usually pulmonary though it is sometimes due to a sound propagated from the aortic orifice

The pulmonary second sound is accentuated normally during youth otherwise it is evidence of abnormally increased pressure in the pulmonary artery and may be due to some obstruction to the flow of blood in the lungs such as emphysema fibroid phthisis or pulmonary arteriosclerosis or to faulty circulation in the left heart

Changes in the Number of the Heart Sounds Instead of the first and second sounds of the heart there may be heard either in healthy or in diseased hearts three or four or even more sounds The additional sounds may be due (1) to failure of the components of the first or second sounds to fuse to a single sound especially when there is a disturbance in the synchronism of events in the two ventricles or (2) to the formation of new sounds

Splittings and Doublings of the Heart Sounds (3/4 Rhythm) The first sound or the second sound is said to be split or doubled when there is no marked alteration in accentuation or in relation to the short and long pauses of the cardiac cycle When the first sound is split there is a slight fault in coincidence When it is doubled the fault in coincidence is greater Similarly the second sound may be either split or doubled Every degree of transition from slight splitting to distinct doubling may be met with These phenomena are frequently noted in health at different phases of respiration The doublings are common too in disease conditions and are then audible in all respiratory phases When the mitral valve is damaged the second sound is often split at the base A splitting of the second sound at the apex is common in mitral stenosis

Triple or Gallop Rhythms (3/4 Time) The normal rhythm of the heart is fairly well maintained in the splittings and doublings Sometimes the heart sounds however appear as groups of three sounds of which the first and second and the second and third are separated by approximately equal intervals while the last member of each group is separated from the first member of the next succeeding group by a somewhat longer pause and for this reason it is fancied to resemble the sounds of the hoofs of a galloping horse (an awkward nag) three hoofs down followed by the fourth hoof (gallop rhythm)

The terms presystolic gallop and protodiastolic gallop are contradictory These extra sounds are related to the complex of sounds constituting the first heart sound They are neither diastolic nor systolic in time Sometimes the extra sound precedes the first sound of the series (presystolic gallop) sometimes it follows (protodiastolic gallop) Gallop rhythms are usually most distinctly audible just medial from the apex of the heart though the rhythm can usually be heard over the whole cardiac area

The existence of gallop rhythm can be confirmed by palpation inasmuch as the extra tone is accompanied by a distinct shock palpable at the apex The tactile perception of the rhythm may be more definite than the acoustic perception

A gallop rhythm may be a normal phenomenon when third sound is audible

It may be heard when a hypertrophied left ventricle is beginning to yield to the strain and therefore its recognition may be more important for diagnosis and prognosis than that of a heart murmur

HEART MURMURS The heart sounds normal or abnormal and heart murmurs are noises never pure tone

The chief difference between the sounds in the normal and the abnormal heart and the heart murmurs lies in the duration and in the termination of the noises The normal sounds are briefer and they cease more quickly Murmurs tend to be longer and to fade away gradually These differences correspond to the origin of the sounds The normal sounds arise from a single sudden disturbance of the equilibrium of the sound producing body whereas heart murmurs are due to a repeated disturbance

In studying heart murmurs attention is paid to (1) the time (or phase) in the cardiac revolution in which the murmur occurs (2) the site at which the murmur is best heard and (3) the direction and propagation of the murmur Less important features are (4) the intensity of the murmur (5) its pitch and (6) its quality or timbre

Timing The first determination to be made is the phase or the phases in the cardiac cycle in which the murmur is audible All murmurs occurring between the beginning of the first sound of the heart and the end of the short pause marked by the beginning of the second sound are termed systolic murmurs All murmurs audible during the period extending from the moment at which the second sound begins to the end of the long pause marked by the beginning of the first sound are diastolic murmurs

The so called presystolic murmur occurring just before the first sound of systole is in reality a diastolic murmur It is close to the normal systolic sound and far removed from the normal diastolic sound

In fetal rhythms when one of the heart sounds ceases to be audible considerable difficulty may be experienced in timing a murmur The best that can be done is to be guided by simultaneous auscultation and palpation of the apex beat or of the carotid pulse

In describing the position of a murmur it is well to state where it is best heard and the direction of its conduction of propagation

Propagation As a rule a murmur is best conducted in the direction of the flow of the current that produces it It is desirable to ascertain not only the point at which a heart murmur is maximal but also the manner in which the murmur is propagated in one or more directions from this point If for example a systolic murmur is produced by narrowing of the aortic valves (aortic stenosis) the murmur will be maximal at the auscultation site for that valve in the second intercostal space to the right of the sternum and the murmur will be propagated upward toward the carotid and the subclavian arteries since this is the direction in which the blood flows from the narrowed spot at which the murmur arises

If the aortic valves leak so that blood can pass back through them into the left ventricle during diastole (aortic regurgitation) the diastolic murmur will be propagated downward and to the left along the left margin of the sternum corresponding to the direction of the regurgitant flow

In narrowing of the mitral valve (mitral stenosis) the blood is forced through a narrow slit directly toward the apex of the heart and during the period of the contraction of the atrium a systolic murmur in the region of the apex is heard If the mitral valve leaks and blood regurgitates through it during contraction of the ventricles (mitral regurgitation) the murmur may be best propagated toward the apex of the heart for instance in a direction opposite to the regurgitation The murmur is sometimes best propagated toward the left atrium and a systolic murmur may be audible either at the apex or in the pulmonary area

When multiple heart murmurs coexist a determination of the maximal and minimal points for each murmur and of the direction in which it is propagated is desirable

If a murmur should be heard equally strongly in two areas it is probable that an autochthonous murmur exists at each site since a murmur propagated any distance is likely to be lessened in intensity

In the presence of coexisting systolic and diastolic murmurs over the heart the origin of the diastolic murmur is first located and analyzed for diastolic murmurs *are likely to be the most significant from the standpoint of diagnosis* Subsequently the systolic murmurs may be analyzed for themselves

As a general rule aortic murmurs are propagated toward the upper part of the body and mitral murmurs toward the lower

Intensity Diastolic murmurs are usually less intense than systolic though not always The two factors concerned in intensity are (1) the size of the opening and (2) the force or velocity of the flow Care should be taken not to overestimate the importance of intensity for prognosis for a very loud murmur may accompany a slight lesion whereas a very soft murmur may coexist with a serious lesion of the heart With a given murmur a patient s condition may be better when the murmur *is intense than when it is feeble since the intensity may vary with the force of the* cardiac contraction

The intensity of a murmur increases and diminishes with the changes in blood pressure When a heart hypertrophies the murmurs in it may become intensified and when a heart weakens the murmurs may grow feebler

The intensity of heart murmurs may also be influenced by posture Most murmurs are more intense when the patient is in the recumbent position than when standing the heart rate is less and the ventricles contract more energetically

The condition of the walls of a diseased orifice may affect the intensity of the murmur produced there Rigid or calcified valves may emit loud sounds whereas soft fresh thickenings of the valves may give rise to feeble murmurs

Quality According to the peculiar quality that heart murmurs manifest they are designated as blowing rasping scratching filing and musical along with any other descriptive adjective the observer may desire to employ

Origin Under normal conditions the relations that exist among the cavities of the heart the valvular orifices the velocity of flow and the composition of the blood are such that no murmurs arise but an abnormal condition of the orifices or a *change in the velocity of flow or in the composition* of the blood gives rise to murmurs

Significance A murmur audible over the heart may arise inside the heart (intracardiac murmur) or outside the heart (extracardiac or exocardial murmur) A murmur may be produced by organic disease of the heart valves (*organic murmur*) or it may be independent of such disease (functional heart murmur) or it may have its origin in conditions not due to disease of the heart muscle or its valves (*accidental murmur*) *in which it may* depend on changes in the physical state of the blood (anemic murmur) or on increased rapidity of flow (velocity murmurs in fevers) or on causes unknown

Organic Intracardiac Murmurs Pathologic changes in the cardiac valves may lead to imperfect closure of the orifice and this is designated valvular *insufficiency* A permanent narrowing of the orifice is known as valvular stenosis Murmurs due to insufficiency of a valve will be produced in those phases of the heart action during *which the orifice is normally* closed whereas murmurs due to stenosis will arise when the orifice is open and the blood current is passing through it in the normal direction

Functional Heart Murmurs Murmurs are frequently audible over the heart during life though at necropsy no changes in the valves of *the heart* are demonstrable

Such murmurs independent of anatomic lesions of the valves are called functional murmurs That a certain number of these are due to imperfect contraction of the muscle ring around the valve leading to a relative insufficiency of the valvular closure has been postulated Relative insufficiencies if such exist are common at the mitral and tricuspid orifices They occur occasionally but rarely at the aortic orifice and perhaps also though still more rarely at the pulmonic orifice (Graham Steell murmur)

In all forms of anemia especially in uncontrolled pernicious anemia systolic murmurs may be heard over the whole heart occasionally an anemic diastolic murmur is audible Perhaps some of the so-called accidental murmurs audible in cachexias may depend on the accompanying anemia

In conditions in which the action of the heart is excited (fevers neurasthenia hyperthyroidism) systolic murmurs are often audible in the pulmonary region and over the left ventricle

A certain number of murmurs have their origin in the lungs during movements of the heart (cardiorespiratory murmurs)

The criterion for differential diagnosis between these less important murmurs and the more important murmurs due to organic disease of the valves or to relative insufficiency is not definite In general the less important murmurs are systolic rather than diastolic They are less precisely localizable than the organic murmurs Many of them are heard in regions in which organic murmurs are rarely present

The less important murmurs are more variable in intensity than organic murmurs especially under the influence of the respiratory movements and of change in posture Most important of all they are not accompanied by those other changes in the heart and circulation that follow an organic disease of the valves

Dorsal Auscultation According to Lian and Dang van Chung diagnosis of certain cardiovascular disturbances cannot be established by auscultation of the anterior aspect of the thorax but may be elicited by auscultation of the back of the thorax This applies particularly to congenital stenosis of the aortic isthmus in which a loud systolic murmur may be heard at the left omovertebral space A systolic murmur can be heard along the left border of the vertebral column in the two inferior thirds of the left side of the thorax in cases of aneurysm of the descending aorta Dorsal auscultation may be an aid in the detection of continuous murmurs due to the compression of a branch of the pulmonary vein or to the presence of an intrapulmonary arteriovenous aneurysm An anterior mitral systolic murmur which is propagated to the back and is heard on dorsal auscultation is highly indicative of mitral insufficiency According to the site of its greatest intensity a systolic murmur at the base of the heart when propagated to the back and detected on dorsal auscultation may be of aid in the diagnosis and the localization of the causative lesion in the aorta (right supraspinous fossa) or in the pulmonary artery (left supraspinous fossa) The continuous murmur caused by the compression of the superior vena cava and that due to patent ductus arteriosus are propagated to the back only during systole Pericardiac friction sounds may be heard on dorsal auscultation in exceptional cases

THE IMPORTANCE OF CARDIAC AUSCULTATION The importance of cardiac auscultation has been recently emphasized by Samuel A Levine who observes that if the competent examiner fails to hear any murmurs whatever even with the patient turned to the left lateral and dorsal position or sitting upright it can be safely assumed that subacute bacterial endocarditis or chronic rheumatic valvular heart disease is not present If a patient who has had rheumatic fever is examined one or more years afterward and fever and murmurs are not observed the heart sustained no important permanent damage However if in a patient who has had rheumatic fever there is any kind of murmur heart disease should be suspected If a diastolic murmur is present in any patient the presence of organic valvular

disease must be suspected or at least cannot be dismissed for auscultation is the only method of detecting early valvular heart disease Likewise the detection of pericardial friction almost invariably means acute pericarditis and this may be first demonstrated by auscultation

In regard to particular valvular heart lesions Levine calls attention to the fact that the presence of an apical rumbling murmur diastolic or presystolic in time is diagnostic of mitral stenosis This murmur establishes the correct diagnosis even if it is faint or present only after effort or when the patient is lying on the left side A faint early diastolic murmur at the base of the heart is most significant in the diagnosis of early aortic insufficiency In some instances auscultation is the only method which will aid in the diagnosis of either slight mitral stenosis or aortic insufficiency for the roentgenograms and electrocardiograms may reveal essentially normal conditions and even symptoms of heart disease may be absent or equivocal

Emphasis in this discussion by Levine is in accord with that of other cardiologists that there are many systolic murmurs of which proper evaluation may be extremely difficult It is well known that systolic murmurs may be heard in both valvular and nonvalvular disease of the heart and may be present in patients who are suffering from noncardiac disease or no disease at all This does not detract from the value of auscultation In general a faint systolic murmur may be heard with or without heart disease but louder ones will seldom be present in normal persons and usually will indicate some form of heart disease

In a patient who has a severe epigastric pain of sudden onset in whom auscultation of the heart reveals a diastolic gallop electrocardiographic studies usually will confirm the suspicion that the heart is the origin of the illness The detection of a snapping first sound while the heart rate is very rapid and grossly irregular in the absence of any murmurs is enough to arouse suspicion of the presence of mitral stenosis Later when the rate slows after digitalis therapy the characteristic diastolic murmur may become audible A hyperactive first sound is present in thyrotoxicosis An exact doubling of the heart rate from 70 to 140 after brief effort arouses the suspicion of the presence of auricular flutter

Many patients who display perfectly good or even loud first heart sounds are observed to have various forms of advanced cardiac disease even with gross congestive failure Others on the contrary who have faint and occasionally inaudible first sounds are healthy

Levine has expressed the belief that in complete heart block when the ventricles are beating very slowly and regularly and the auricles more rapidly regularly and independently the first sound varies greatly in intensity It is found to be loudest when P R interval is exceedingly short and often the observer may be able to estimate the length of the P R interval and to recognize conditions in which there is a short P R interval others with a prolonged P R interval (first degree heart block) and those in which this interval is varying such as second and third degree heart block In Levine s opinion the detection of these changes in the intensity of the first sound is the only reliable bedside method of making these diagnoses If the heart rate is regular and the first sound decidedly decreased while the second sound is normal the P R interval will be at least at the upper limit of normal or delayed systole Similarly if the rate is regular and slow and the first sound changes in intensity with sudden loud explosive sounds there is complete heart block with dissociation of the auricles and ventricles

Exocardial Murmurs Abnormal sounds audible over the heart synchronous with its action but arising outside its cavities are (1) pericardial friction (see Pericarditis) (2) pleuropericardial friction (3) cardiorespiratory murmurs (4) precordial crackling characteristic of mediastinal emphysema (see Diseases of Mediastinum Chapter 9) and (5) splashing sounds

Pleuropericardial Friction Sounds A pulsatile friction sound produced by rubbing of the outer surface of the pericardium against the pleura may be synchronous not only with the heart s action but also with the respiratory movements The portion of the sound due to the latter will cease when the breath is held

Cardiorespiratory Murmurs These sounds arise in the lungs chiefly during the systoles accompanying inspiration Occasionally diastolic sounds are produced and sometimes the sounds are audible during expiration also The heart becomes smaller during systole and the ingress of air in the adjacent lung is increased owing to greater negative pressure in the lung The sounds may be blowing like those of vesicular breathing or crackles and rales may be produced The systolic vesicular breathing may closely resemble a heart murmur These pulsatile pulmonary sounds usually cease or are markedly modified when the breath is held They are also much influenced by changes in posture

When the lung is adherent in front of the vessels at the base of the heart a diastolic murmur sometimes becomes audible the diastolic retraction of the aorta causing a localized aspiration into the adjacent pulmonary alveoli

Splashing Sounds If the air and fluid occur together in the pericardial cavity each beat of the heart may give rise to a metallic ringing splash Similar sounds are sometimes heard when there is a pulmonary cavity near the heart when a hydropneumothorax exists or even when the stomach is much distended with fluid and gas

If sounds have an extrapericardial origin they will disappear when the patient is sitting to reappear when he lies down but if they have an intrapericardial origin the sound is heard in both positions

Auscultation of the Blood Vessels When one is listening over the blood vessels care must be exercised not to exert pressure with the stethoscope otherwise a narrowing of the vessel will be produced which will give rise to a murmur

Auscultation of the Arteries The observer locates the vessel by palpation and sets the bell of the stethoscope lightly upon it Normally two sounds are audible in the subclavian and the carotid arteries the first just after the beginning of ventricular systole the second just after the beginning of ventricular diastole If the artery is pressed on now with the stethoscope a ventriculosystolic stenosal murmur will become audible No sounds are audible normally over the other arteries of the body

Systolic murmurs due to narrowing at the aortic orifice are often propagated into the carotid and the subclavian arteries When the aortic second sound is absent as in some cases of aortic insufficiency no arteriosystolic tones are heard in the great vessels

When a Corrigan pulse exists the pulse wave rising quickly and sinking again very rapidly murmurs are often audible in the arteries of the neck and in the femoral and the brachial arteries The phenomenon is most often met with in aortic insufficiency but it may also be encountered in hyperthyroidism in fever and in nervous palpitation When a very loud sound is audible it is spoken of as a pistol shot sound

In aortic insufficiency on listening over the femoral artery two tones quickly following one another are sometimes to be heard (Traube s double tone) On slight pressure with the stethoscope these tones disappear and first is heard a normal pressure murmur and later on stronger pressure a second murmur (Duroziez s double murmur) Duroziez s double murmur is hard to differentiate from the normal pressure tone

Any compression of an artery may give rise to a stenosal murmur for example this fact is utilized in the auscultatory method of determining maximal and minimal blood pressure

Auscultation of the Right Jugular Vein The stethoscope is placed over the sternal attachment of the right sternocleidomastoid muscle the patient sitting or standing with the head turned toward the left

In the jugular vein certain sounds become audible under abnormal conditions they include (1) the venous hum and (2) the venous tone

In anemic patients and occasionally in healthy persons a continuous blowing singing or humming murmur with systolic accentuation heard over the jugular vein is a venous hum Less continuous more intermittent venous murmurs are unimportant

Venous tone is occasionally audible in the same situation in tricuspid insufficiency it is due to sudden tension of the dilated vein when the blood propelled by the contracting right ventricle rushes into it

Movements of the Heart and Blood Vessels The signs of the activity of the heart include the heart sounds (page 644) and certain movements of the heart and blood vessels that are accessible to clinical examination These movements are visible and palpable as pulsations in the heart and its neighborhood and in the arteries and veins of the body The movements can be most accurately analyzed by means of instruments of precision yielding graphic records

With the hand most of the movements which can be seen can be felt and a number of movements that cannot be seen are recognizable by *palpation* Thus for example an apex beat that is invisible may sometimes be felt various invisible shocks and thrills are palpable and in addition details of the movements of the blood vessels that are not accessible to inspection can be made out on palpation

In practicing palpation the palm of the hand (the tips of fingers for the pulse) is applied lightly and with varying degrees of pressure on the area to be examined The *region of the apex of the heart first and successively the* whole precordial area the axillae the vessels in the neck the epigastric and hepatic regions and the more superficial vessels in various parts of the body including of course the vessels of the feet are palpated

Low pitched murmurs are the ones that favor the formation of thrills whereas those that yield rapid vibrations producing high pitched murmurs may not give rise to palpable thrills This explains why a thrill may sometimes be felt when a murmur is not audible or is a very indistinct rumble and why the loudest and most distinct blowing murmurs may be unaccompanied by thrills Palpation and auscultation here supplement one another advantageously in diagnosis

Abdominal Pulsations Pulsation due directly or indirectly to the heart is present in the epigastric fossa in the neighborhood of the xiphoid process between the two costal margins It may depend on (1) the abdominal aorta or (2) the right ventricle

The commonest aortic form is that met with in neurasthenics and emaciated dyspeptics with thin loose abdominal walls the so called dynamic aorta This pulsation is slightly to the left of the middle line and extends for a variable distance downward below the xiphoid process The pulsation is perpendicular from behind forward has only a slight breath and is somewhat later than the apex beat One tone or a systolic murmur may be audible on auscultation The pulsation is increased by anything that excites the heart s activity In many cases in which this form of aortic pulsation is present the aorta can be grasped in the palpating hand which then becomes aware of marked lateral expansion with each pulsation In such cases however the aorta can be palpated lower down and the same condition found there is never a localized expansile tumor such as is present in aneurysm of the abdominal aorta

Epigastric pulsations due to the right ventricle itself may be either systolic elevations or systolic retractions Systolic elevations are usually due to a lowered

and enlarged right ventricle (pulmonary emphysema dilated right heart right heart failure cardioptosis)

Systolic retractions in the epigastrium due to the right ventricle are rather diffuse wavelike movements depending on elevation of the diaphragm by the contracting right ventricle they are of no clinical significance except when there is failure of the right side of the heart

VISIBLE PULSATIONS Visible pulsation of the peripheral arteries occurs in aortic insufficiency For the characters of the arterial pulse however reliance is made on palpation

Pulsations in the veins of the neck may be visible in health though they are much more often seen in disease A normal venous pulse when visible presents two waves recognizable by the eye both diastolic Palpation is of little value in the study of the venous pulse

A visible capillary pulse is met with in conditions associated with hypertrophy of the left ventricle especially in aortic insufficiency If a line is scratched with the fingernail on the forehead or on the skin of the trunk or if light pressure is made on the end of the patient s fingernail in order to make a pale spot in the nail bed the borders of which may be closely watched alternately blush and pallor synchronous with the pulse are revealed if a capillary pulse exists A very good way to look for a capillary pulse is to press gently with a glass slide on the lips Sometimes a blush of the cheek can be seen with each systole of the heart

THE PULSE By pulse is meant the pressure wave of enlargement of the artery which occurs at each systole of the heart and which takes a perceptible time to travel from the heart to the periphery Attention is paid to the following qualities (1) the frequency (2) the rhythm (3) the volume (4) the quickness or celerity (5) the tension and (6) the equality on the two sides of the body

The thickness of the vessel which is usually attended to at the same time is not a pulse phenomenon but has to do with the condition of the arterial wall itself

Rate This varies in healthy adults between 60 and 90 beats per minute in children 90 to 140 in old age 70 to 90 The pulse is faster in women than in men On sitting or on lying the pulse is slower than on standing or on exercising

Acceleration of the pulse rate is due to cardiac hurry or tachycardia It is met with normally on exertion during emotion pain fever and after taking food

In hyperthyroidism and in some neurasthenic states tachycardia is common A frequent pulse is often an important sign of cardiac weakness or collapse In paroxysmal tachycardia attacks of great frequency of rate having a sudden onset and ceasing suddenly are met with alternating with periods of normal frequency The attacks may last from a few minutes to several days the pulse beats often *numbering between 140 and 280 per minute*

Slowing of the pulse rate is known as bradycardia It is met with in convalescence from many infectious diseases especially typhoid fever influenza and pneumonia in disturbances of digestion in conditions in which the vagus centers are stimulated (brain tumors hydrocephalus beginning meningitis) in icterus and in various diseases that affect the heart itself (aortic stenosis coronary sclerosis myocarditis) In Adams Stokes syndrome in which the stimulus from the sinus and auricles is prevented from reaching the ventricles the latter contract in their own independent rhythm in these cases the arterial pulse rate may fall below 30

Rhythm Normally the single pulse waves follow one another regularly but in pathologic conditions the rhythm may become irregular The irregular pulse is due to cardiac arrhythmia Under this heading there are included (1) respiratory irregularities (2) extrasystolic irregularities (3) heart block (4) perpetual arrhythmia and (5) the alternating pulse

Volume What clinicians speak of as the volume of the pulse is dependent chiefly on the difference between the increase in pressure during ventricular systole and

the decrease in pressure during ventricular diastole The size of the pulse waves depends chiefly on (1) the volume of the systolic output of the left ventricle and (2) the ease with which blood flows out of the arteries through the capillaries The volume of the pulse is therefore the palpatory equivalent in the radial artery of what is known as the pulse pressure (difference between maximal systolic and minimal diastolic pressure)

If the pulse wave rises very quickly and falls rapidly it is spoken of as a water hammer pulse (Corrigan pulse)

Tension This is judged by the force required to obliterate the pulse when the fingers press on it Three fingers are placed on the radial artery the most pressure is applied with the distal finger hard enough to prevent a recurrent pulse wave through the palmar arch pressure is then made with the most proximal finger until the pulse ceases to be perceptible to the finger in the middle If difficult to compress the pulse is said to be of high tension when easily compressible it is of low tension

Equality The commonest cause of differences in the pulse in the two radial arteries is the presence of an abnormally small radial artery on one side This is a common anomaly in the arterial system of the forearm As a result the pulses vary chiefly in absolute volume they are equal in time In aneurysm of the arch of the aorta or of one of the arterial trunks supplying the arms differences in the time as well as in the volume and tension of the two pulses are prominent

Changes in the Cardiac Rhythm The clinical significance of changes in the cardiac rhythm especially the cardiac arrhythmia has been greatly clarified by the studies of Thomas Lewis and by the use of electrocardiograph Increase in the cardiac rhythm beyond the limits of the normal number of heart beats per minute is tachycardia Decrease in the cardiac rhythm below the limits of the normal number of heart beats per minute is bradycardia

The *tachycardias* with a *regular pulse* rhythm as proved by the electrocardiogram include sino auricular tachycardia paroxysmal auricular tachycardia ventricular tachycardia and most instances of auricular flutter Tachycardias with an *irregular pulse* rhythm include auricular fibrillation and sinus tachycardia with premature beats

Bradycardias with *regular pulse* rhythm include sinus bradycardia nodal rhythm constant partial heart block and complete heart block *Irregular bradycardias* include *sino auricular bradycardia with frequent* premature beats and partial heart block with frequent dropped beats

Irregular rhythms with normal heart rate are found to include premature beats auricular fibrillation with ventricular rate slowed by digitalis sinus arrhythmia and partial heart block with dropped beats

In *sinus tachycardia* the cardiac impulse as recorded on the electrocardiograph arises normally in the sino-auricular node The heart rate is increased Since the heart rate of normal individuals varies greatly it is well not to interpret an increased pulse rate as being abnormal until it is known whether the particular rate is abnormal for the patient in question The origin of the activating impulse is revealed by the electrocardiograph If the impulse arises in a ventricular focus it causes ventricular premature beats If the impulse arises above the bifurcation of the bundle of His it causes supraventricular premature beats In supraventricular premature beats the impulse arises in the auriculoventricular node and thus the beats are termed nodal premature beats If the impulse arises elsewhere in the atria there are auricular premature beats

In tachycardias there may be no subjective symptoms or there may be disagreeable palpitation fatigue and breathlessness

Examination of the heart and pulse discloses a regular rhythm with a rate exceeding 90 to 100 but less than 160 per minute *Differentiation from other tachycardias with regular rhythm may be surmised or made from the electrocardiogram*

In *sinus bradycardia* the cardiac impulse arises normally in the sino auricular node as revealed by the electrocardiograph Symptoms of sinus bradycardia are absent except in the extremes when there is dizziness faintness or syncope Syncopal attacks if they occur may be due to cardiac asystole Examination of the heart and pulse discloses a regular rhythm with slowing of the heart rate below 60 per minute

In *sinus respiratory* or *juvenile arrhythmia* the cardiac impulse arises normally in the sino auricular node as revealed by the electrocardiograph and the arrhythmia is characterized by alternating periods of slower and more rapid cardiac rates which are usually related to respiration The heart rate increases with inspiration and diminishes with expiration

Sinus arrhythmia is recognized clinically by regular but more rapid heart beats during inspiration and slower beats during expiration The periodic variation in the rate is intensified by deep breathing and stopped by breath holding or by exercise A sinus arrhythmia is of no clinical importance

In *sinus arrest* (sino auricular standstill or sinus pause) there is momentary failure of the sinus node to initiate an impulse The failure is caused usually by digitalis or quinidine intoxication Clinically the rhythm is regular but there is slowing of the pulse An electrocardiogram is essential for diagnosis

When heart beats are controlled by the *auriculoventricular node* there is a shifting pacemaker Auriculoventricular rhythm occurs relatively infrequently It is almost always a transient phenomenon alternating with sinus rhythm Clinical symptoms are uncommon and rarely distressing Diagnosis requires electrocardiographic study

When there is shifting of the pacemaker there may be *reciprocal beats* that is there is an auriculoventricular rhythm in which the ventricles contract before the atria The atrial activation is delayed long enough so that the ventricles can respond to the returning auricular (nodal) impulse This phenomenon is often induced by pressure on the carotid sinus or digitalis therapy Diagnosis is by electrocardiogram

In other instances of a shifting of the pacemaker there may be *auriculoventricular rhythm* and *auriculoventricular dissociation* In auriculoventricular dissociation there are responses to different pacemakers The ventricular rate controlled by the auriculoventricular node may exceed that of the atria which are responding to a depressed sino auricular node This type of auriculoventricular dissociation is usually the result of digitalis or of infections such as rheumatic fever In this condition the dissociation results from complete auriculoventricular block The electrocardiogram is necessary for diagnosis

Premature beats or extrasystoles are cardiac contractions of ectopic origin which occur earlier than expected in the dominant or usual rhythm They are distinguished electrocardiographically by their point of origin for instance auricular auriculoventricular or nodal and ventricular premature beats

When there is recurrently a normal beat followed by an extrasystole the rhythm is coupling of beats (bigeminal pulse) Bigeminal pulse is induced by two beats in close proximity followed by a long compensatory pause The commonest type of coupling is due to alternate extrasystoles and is caused by digitalis Coupling of beats may also result from partial auriculoventricular block not the result of digitalis in which every third beat (trigeminal pulse) is dropped and thus a 3 2 sino auricular block exists If every fourth beat is blocked there is a 4 3 sino auricular block

The majority of patients who have premature beats do not have organic heart disease The presence of heart disease may increase the occurrence and frequency of premature beats In those who have premature beats and do not have heart disease emotional stress and conflict mental or physical fatigue irregular sleep and digestive disturbances especially when these are combined with excessive smoking of tobacco or drinking of alcohol or coffee increase the beats

Premature beats occur at all ages After the age of 50 years the frequency in creases and after 70 years premature beats are very commonly present After the patient is aware of premature heart beats there may be a sensation of stoppage of the heart momentary faintness or dizziness and anxiety The patient may describe palpitation as an oppression fluttering thumping skipped beats, or a sinking feel ing Occasionally the discomfort is localized in or extends to the neck where the patient experiences a sensation of fullness tightness pulsation or wave reaching to the top of the head This may be caused by the regurgitant stream from the right atrium to the veins of the neck when the atrium contracts simultaneously with th ventricle and the tricuspid valve is closed

During the occurrence of a series of extrasystoles there may be anxiety pallor sweating nausea weakness dizziness faintness and breathlessness

Premature beats often become annoying when the patient retires for the night Conversely exercise or anything which increases the heart rate eliminates the pre mature beats

Premature beats are recognized by auscultation occasionally by palpation of the radial pulse They may be absent at the time of physical or electrocardiographic examination The electrocardiogram is essential for definite diagnosis and localiza tion of premature beats

Auricular paroxysmal tachycardia is manifested by a rapid but regular heart rate The consensus is that paroxysmal tachycardia is due to a special kind of circus rhythm (re entry) which involves the sino auricular node or the auriculoventricular node in its pathway

The etiology of auricular paroxysmal tachycardia is obscure but it seems that organic heart disease is usually absent Auricular paroxysmal tachycardia occurs at any age including infancy It is commonest between 20 and 40 years of age

Paroxysms of brief duration may be accompanied by mild palpitation or no symptoms at all In the severer attack there is sudden onset with a thump or two against the thoracic wall followed by cutaneous palpitation fluttering racing or pumping of the heart beat Often there are stenocardia and smothering sensations or epigastric distress

The psychic and reflex nervous manifestations may be profoundly expressed by angor animi syncope anxiety weakness exhaustion coldness sweating dizziness and faintness Abdominal distention gaseous eructations salivation nausea or vomiting or polyuria may terminate the attack

It is remarkable how often heart rates of more than 200 per minute in parox ysmal tachycardia are tolerated for hours or days without evidence of circulatory insufficiency As a rule the paroxysm ends abruptly normal rhythm is restored and the symptoms and signs of circulatory insufficiency abate rapidly In hearts in which there is organic disease it is recorded that prolonged tachycardia induces failure

In the extreme and very exceptional instance the attack is prolonged In such case there may be peripheral slowing of the circulation to the extent that thrombosis and gangrene of an extremity occur The attacks can terminate fatally either with signs of progressive failure or suddenly

On physical examination the only abnormality is a regular and constantly rapid heart rate In those who have organic disease there may be cardiac enlargement and murmurs

In most cases auricular paroxysmal tachycardia is of no clinical importance either from the viewpoint of producing disturbing symptoms or from that of de noting the presence of organic heart disease except in very rare instances in which the paroxysms recur frequently or are very prolonged

The most useful therapeutic and often diagnostic procedure is to make pressure over the right carotid sinus and if this is ineffective over the left carotid sinus for

10 to 30 seconds The carotid sinus may be located by feeling the strong carotid arterial pulsations at the bifurcation of the common carotid artery at the level of the upper border of the thyroid cartilage The patient may be reclining or sitting with his head slightly forward and his neck relaxed Pressure is best made with two or three fingers compressing the artery and sinus posteriorly and medially against the vertebral spine Occasionally a massaging motion is effective Prolongation of the procedure is avoided because of the possible risk of syncope or cardiac standstill

Auricular flutter is due to a mechanism related to auricular fibrillation or auricular tachycardia—a circus rhythm

In auricular flutter the atria beat regularly at a rate of 250 to 350 per minute As a rule however there are varying degrees of partial heart block (2 1 3 2 3 1 and others) Most commonly the ventricular rates vary between 100 and 150 per minute and there is a 2 1 block With higher degrees of block for example 4 1 the ventricles may beat not only regularly but at normal rates 70 to 80 In infants and rarely in adults there is a 1 1 ventricular auricular rate

Auricular flutter does not necessarily indicate heart disease However it is often associated with mitral stenosis and it may be present with hyperthyroidism coronary arteriosclerotic disease and hypertensive heart disease Auricular flutter may be associated with mediastinal disease and may occur during surgical procedures in the thoracic cavity

The symptoms of auricular flutter are sudden tachycardia palpitation fear of impending death weakness dizziness and syncope and in some cases there are the symptoms and signs of congestive heart failure or shock

Auricular flutter is often more disturbing to the patient than auricular tachycardia because of its greater tendency to persist (days weeks months or years) Auricular flutter is likely to be followed by auricular fibrillation In auricular flutter the pulse is regular

Auscultation of the heart discloses apical rates which depend on the degree of heart block (2 1 3 1 or 4 1) Sudden changes in rates indicate changes in the degree of the block Exercise may raise the ventricular rate and induce a constant degree of block and a regular heart beat

The diagnosis is established by means of electrocardiographic studies

Auricular fibrillation is an irregular irregularity of the heart beat Like auricular flutter it is regarded as a circus rhythm of an ectopic stimulus In auricular fibrillation however the basic circuit is shorter and the frequency of circulation of the excitation wave is greater than in flutter

Auricular fibrillation is associated with various degrees of auriculoventricular block The auricular rhythm is irregular the ventricular rhythm also in auricular fibrillation is irregular

Auricular fibrillation is commonly associated with severe organic heart disease It may be present however in patients who have apparently otherwise normal hearts Auricular fibrillation occurs most commonly in rheumatic heart disease coronary arteriosclerotic heart disease with heart failure and hyperthyroidism Paroxysmal auricular fibrillation is particularly identified with hyperthyroidism although it may occur in any type of heart disease including congestive heart failure as well as subacute bacterial endocarditis It may occur in paroxysms in the first two weeks after an acute myocardial infarction but in the latter condition it usually subsides spontaneously after several hours Most frequently auricular fibrillation is observed in patients more than 40 years old Women are more commonly affected in the younger age period that is in hyperthyroidism and mitral stenosis men in coronary artery disease and hence in the older age periods

Often the onset of permanent auricular fibrillation impairs the cardiac sufficiency to a degree that heart failure is chronic thereafter In any patient who has auricular

fibrillation auricular thrombosis may occur However thrombus formation and subsequent infarctions are common in mitral stenosis with or without auricular fibrillation

The heart rate and the pulse rate possess the quality of irregular irregularity In rapid ventricular rates the pulse rate is considerably less than the apical rate this difference is termed a pulse deficit The pulse deficit results from failure of many of the weak ventricular beats to expel a sufficiently large column of blood to reach the radial artery The pulse deficit is increased by the inability of the counter to count weak pulse beats which are closely spaced

In those individuals who have pulse deficits the diagnosis is made by this finding In all cases electrocardiographic studies are necessary for a diagnosis

In an otherwise normal heart paroxysmal auricular fibrillation may be disturbing but it is usually of no importance as regards the outlook for life or the development of heart disease

In the presence of organic heart disease a persistent auricular fibrillation is an unfavorable occurrence

Treatment for a *paroxysm* of auricular fibrillation is indicated only if the paroxysm is associated with distressing and prolonged palpitation evidences of heart failure or of cerebral or peripheral ischemia

In all other instances if treatment for auricular fibrillation is necessary it consists essentially of the use of digitalis or quinidine

Ventricular tachycardia is a rapid regular rhythm initiated by stimuli arising in an ectopic focus in the ventricular conduction system The ventricular rate varies usually between 150 and 300 per minute The atria may contract regularly and normally or may respond to retrograde conduction along the auriculoventricular bundle and node while the ventricles respond to the ectopic ventricular stimulus

Paroxysmal ventricular tachycardia is most often associated with recent myocardial infarction It may occur without myocardial infarction in association with hypertensive and arteriosclerotic heart disease after excessive exertion and after excessive administration of digitalis Occasionally however ventricular tachycardia is observed without evidence of organic heart disease

The symptoms of ventricular tachycardia are identical with those of auricular paroxysmal tachycardia The diagnosis depends on the electrocardiographic studies

Ventricular tachycardia is a serious condition Ventricular tachycardia with alternating direction of the ventricular complexes has been considered especially ominous and almost invariably fatal In some patients who do not have organic heart disease other than the arrhythmia the prognosis is good Administration of quinidine constitutes the treatment

Ventricular fibrillation is observed as the result of myocardial infarction general anesthesia lightning shock digitalis or quinidine and as a terminal event in a variety of diseases It is likely due to an inconstant circus rhythm

Ventricular fibrillation probably occurs more frequently in those who have not had heart disease than in those who have had it for it is the terminating event in the lives of many individuals In heart disease it is the terminating event in angina pectoris and in acute coronary occlusion and syphilitic aortitis The intravenous injection of drugs is often responsible for this serious cardiac arrhythmia Faintness occurs when ventricular fibrillation lasts 3 seconds syncope after 10 to 20 seconds and convulsions apnea and incontinence after 40 seconds

Electrocardiographic studies are diagnostic if life is sufficiently prolonged to make the diagnosis If ventricular fibrillation is not immediately fatal the patient may recover

Heart block results from aberrations in the sino auricular node auriculoventricular node bundle branch and short P R interval with prolonged QRS complex

Sino auricular block may result from vagal stimulation for instance a sensitive carotid sinus tumors compressing the carotid sinus vagal reflexes during anesthesia and surgical procedures and cerebral lesions The usual cause however of sino auricular block is poisoning by digitalis and occasionally quinidine

There are usually no clinical symptoms unless there are frequent and prolonged pauses However in prolonged pauses there may be dizziness or faintness and even attacks of the Adams Stokes syndrome may occur Death resulting from cardiac standstill is not expected

Sino auricular block is distinguished by the electrocardiogram It is treated by stopping the administration of the heart medicine

In *auriculoventricular block* there is a delay in transmission of the impulse through the auriculoventricular node and bundle or an interruption of an occasional impulse or of all impulses in either the node or the bundle Thus there are variations in degree of heart block from none at all to complete block

Complete heart block is a common disorder of the heart beat appearing in re current attacks often leaving normal conduction between attacks The frequency of heart block is exceeded only by that of premature beats auricular fibrillation and auricular paroxysmal tachycardia

Complete heart block may be transient or permanent and it may represent a functional or an organic disturbance

Coronary arteriosclerosis rheumatic heart disease syphilitic heart disease neo plasms and congenital heart disease and effects of the common drugs (digitalis quinidine) used in therapy are the usual causes of heart block Occasionally heart block is caused by acute infectious diseases notably rheumatic fever and diphtheria but at times by almost every other infection

In most instances of heart block associated with acute infections there is only a prolongation of the P R interval In such cases the P R interval may often be shortened to normal by the administration of atropine

In the normal electrocardiogram the P R interval varies between 0 12 and 0 2 second At slow cardiac rates (60 per minute or less) a P R interval up to 0 21 or 0 22 may be normal In prolonged auriculoventricular conduction time the P R interval exceeds 0 2 second In partial heart block there periodically is a dropped beat The P R interval may increase progressively until a QRS is omitted until the ventricles respond only to every second third or fourth beat resulting in 2 1 3 1 or 4 1 block

There are no symptoms The heart rate increases with exercise or excitement

In complete heart block there is the virtual regularity of atrial and ventricular activity with continual variation in the P R interval The auricular rate is increased over the ventricular rate and both rhythms are essentially regular

In complete heart block the *Adams Stokes syndrome* may occur The Adams Stokes syndrome results from a prolonged systole of 5 to 10 seconds or more As the result the patient becomes pale and pulseless and loses consciousness When the asystole lasts for 15 seconds or more there is cyanosis and respirations are deep and labored or of the Cheyne Stokes type There is a generalized convulsive seizure which lasts for a minute or longer Then the convulsive movements cease and the pulse and respirations return to normal The color is restored Paroxysms longer than a minute may be fatal Recovery may follow longer periods of asystole

Syncope from cardiac standstill may occur in the normal heart as a result of vagal reflexes from a sensitive or compressed carotid sinus or in instances of sino auricular block This is the type of syncope enjoyed by fashionable ladies of former days

In partial or complete heart block the cardiac rate is slow 50 to 40 per minute or less The lower rates prevail in complete heart block that is congenital rheumatic or due to digitalis

In complete heart block variations in the intensity of the first heart sound may

be heard The auricular sound sometimes may be heard faintly between the regular heart sounds or there may be reduplication of the first or second sound which may be thought to be auricular sounds

The clinical significance treatment and the prognosis of heart block depend on the causative factor

In *bundle branch block* there is a delay in or an obstruction to the conduction of impulses through one of the branches of the bundle of His or there are obstructions in the Purkinje fibers

Left and right bundle branch blocks are identified Left bundle branch block occurs more frequently than right bundle branch block

The great majority of instances of bundle branch block (left bundle branch) are due to coronary arteriosclerosis and associated hypertension The bundle branches are often damaged or blocked by cardiac infarction and degenerative changes secondary to myocardial ischemia

There are no symptoms of bundle branch block The definite diagnosis depends on the revelations of the electrocardiogram

Blood Pressure MEASUREMENT OF BLOOD PRESSURE Clinically the arterial pressure in the brachial artery is measured routinely The maximal arterial blood pressure or systolic pressure is *recorded as the highest point reached by the* blood pressure in the artery during the ventricular systole The minimal arterial blood pressure or diastolic pressure is recorded as the lowest point reached by the blood pressure within the artery during ventricular diastole

If the minimal blood pressure is substracted from the maximal blood pressure the remainder is known as the pulse pressure The term mean pressure is used to designate the average pressure *during a certain period*

The principles underlying all the instruments for determination of blood pressure are the same The arterial pulse is obliterated by compression from without by means of a cuff and the pressure necessary for compression is measured by some form of manometer These instruments include the mercury manometers and aneroid manometers The mercury manometers are preferable for office use The aneroid manometer is convenient for bedside use being small and easily portable It may require however to be adjusted at intervals on comparison with a mercury manometer

ARTERIAL BLOOD PRESSURE A cuff is applied over the brachial artery and the pressure in it is raised until the radial pulse disappears The bell of the stethoscope is then placed over the artery close to the cuff the position of the bell is the antecubital space It is customary to speak of five distinct phases as the sound varies during the fall of the pressure in the cuff

First Phase When the pressure falls to the level of the maximal systolic pressure an arterial tone is heard not unlike the first sound of the heart It is due to the return of the pulse wave in the artery the vibrations being supplemented by the resonance of the air within the cuff This is the maximal (systolic) pressure

Second Phase As the pressure in the cuff falls further the sound of the first phase is accompanied by a hissing murmur as the blood flows through the constriction into the wider artery below

Third Phase As the minimal diastolic pressure is approached the murmur of the second phase disappears and a tone usually much louder than that of the first phase becomes audible

Fourth Phase At the end of the third phase if the examiner listens attentively the sound will be observed to become diminished suddenly and markedly and to assume a muffled character The onset of this fourth phase formerly was considered to indicate the exact moment at which the pressure within the cuff corresponds to the minimal (diastolic) pressure in the brachial artery Recently however a committee chosen by the American Heart Association has decided that in clinical practice the recorded diastolic pressure shall be the reading of the manometer at the end of the fif hase

Fifth Phase This is very short at the end of it the sound disappears entirely This is now considered to be the diastolic pressure However this arbitrary standard for diastolic pressure remains with the one limitation of all auscultatory methods It is somewhat unsatisfactory when tones are audible in the peripheral arteries before the cuff is applied (for example in aortic insufficiency)

CLINICAL ESTIMATION OF BLOOD PRESSURE The recorded level of blood pressure in addition to individual fluctuation is affected by numerous technical circumstances such as the nature of the apparatus and whether the measurement is made by auscultation or by palpation In a study of the accuracy of blood pressure recordings Ragan and Bordley compared auscultatory pressure measurements with intra arterial pressure measurements 138 times in 51 adult subjects They found that the agreement between the auscultatory and intra arterial measurements of systolic pressure was affected both by the size of the subject s arm and by the contour of the pulse wave They concluded therefore that the commonly employed clinical method of measuring blood pressure should not be considered a truly accurate procedure and that misinformation is particularly likely to be obtained in subjects who have unusually large or unusually small arms If the arm is small the clinical estimate of the systolic pressure is likely to be too low if the arm is large the clinical estimate of both systolic and diastolic pressures is likely to be too high The error in either direction may exceed 30 mm of mercury The error can be reduced in large armed or fat armed individuals if the width of the cuff is increased as it is in the cuffs for leg pressures In small armed adult persons and in children the error can be reduced by the use of successively narrower cuffs

THE RANGE OF NORMAL BLOOD PRESSURE Master and associates record that the normal range of systolic blood pressure in men starts at the age of 16 years with 105 to 135 mm of mercury and at ages 60 to 64 years reaches 115 to 170 In women it starts at a slightly lower level at 16 years of age the normal range is 100 to 130 mm of mercury At ages 60 to 64 years the range is 115 to 175 This is slightly higher than in men

In men at ages 16 to 18 years the lower limit of hypertension is 145 mm of mercury systolic at 19 years 150 mm and at 30 to 34 years 155 mm It then increases about 5 mm every five years At ages 60 to 64 years 190 mm is the lower limit In women the lower limit of hypertension is 140 mm of mercury systolic up to age 30 years Thereafter it increases 5 or 10 mm for every five years and by age 45 years it is actually higher than in men The hypertensive limit remains higher than among men up to ages 60 to 64 years when it may be taken as the same in the two sexes

The range of normal diastolic pressure begins at 60 to 86 mm of mercury at age 16 years in men and at 65 to 85 mm in women The change with age is gradual By ages 40 to 44 years the level is 70 to 94 mm for men after age 40 years the lower limit of normal remains the same but the upper limit of normal gradually increases to 100 mm at ages 60 to 64 years In women the normal range at 40 to 44 years is 65 to 92 mm and at ages 50 to 54 years rises to 70 to 100 mm it remains at this level to 60 to 64 years of age

Mean blood pressure readings both systolic and diastolic increase with age in both sexes Average systolic readings for men show a fairly smooth progression up to the age of 50 years after that age the increase accelerates For women the increase in the averages by age is somewhat less smooth or continuous than it is for men but among them too it is accelerated after the age of 50 years Beginning with age of 20 years the upward progression in diastolic pressure with age is fairly steady for both sexes systematic acceleration of the average is not found over any broad age period

Among boys between the ages of 16 and 19 years an upward trend is observed in the averages for both systolic and diastolic pressures Among girls on the other

hand values for both systolic and diastolic pressure are practically identical between the ages of 16 and 18 years and remain unchanged through the age period 19 to 24 years Explanation of this difference may lie in the fact that boys mature between the ages of 16 and 19 years they become heavier and are much more active than are girls at these ages Girls mature earlier It has been long recognized that a rapid rise in blood pressure occurs during puberty and adolescence

No clear cut relationship between height and blood pressure has been established

Weight is an important and consistent factor in blood pressure changes With increase in weight there is progressive increase in the averages in systolic and diastolic blood pressure regardless of age or sex

The difference in the mean systolic blood pressure of underweight and over weight groups at each extreme exceeds 10 mm of mercury in some instances the difference in diastolic pressure is almost as great Among women particularly after the age of 40 years the differences between the weight groups at either extreme with respect to the mean pressures both systolic and diastolic are even greater than are those among men Absolute and relative variations in blood pressures as measured by the standard deviation and the coefficient of variation also tend to increase with weight particularly after the age of 40 years In general the greatest contrast in the mean pressure is found at the older ages and among women

Blood Pressure Variations in the Two Arms In 1 388 blood pressure readings Rueger found the blood pressure to be the highest in the right arm in 992 readings as compared to 274 in the left arm and to be equal in both arms in only 122 readings The systolic variation in the two arms was 10 or more points in 694 readings and zero to 10 points also in 694 readings out of the total of 1 388 tabula tions The diastolic variation in the two arms was much less marked with 920 read ings showing less than 10 points difference as compared to 468 readings showing a difference of 10 or more points

Casual and Basal Blood Pressures Smirk defines casual blood pressure as the sum of the basal blood pressure and the supplemental pressure this last r pre sents the degree of blood pressure elevation above the basal level due to whatever degree of physical emotional and suprabasal metabolic activity is present at the time of blood pressure measurement The basal pressure is the pressure measured at a time when physical emotional and metabolic activity are reduced to a physiologic minimum Failure to obtain the true basal reading will not always be apparent to the observer especially when the failure is due to emotional reasons *Both normal and hypertensive subjects with a high basal pressure have a greater statistical ex pectation of having a high casual pressure than do those whose basal pressure is low Likewise those with high supplemental pressures have a greater statistical expectation of a high casual pressure than do those whose supplemental pressure is low* In comparing one individual with another the basal and supplemental pres sures however are independent variables in the sense that the level of the basal blood pressure in an individual is no guide to the probable level of the supplemental pressure *Most statistics concerning the level of the blood pressure are concerned with the casual readings* The fact that when comparing one individual with another within a comparable physiologic group the casual blood pressure is to be regarded as the sum of two independent variables has statistical implications *The supple mental pressure forms a significantly higher proportion of the casual pressure among patients who have essential hypertension than among normal subjects*

Low Blood Pressure (Hypotension) Systolic blood pressure in the range *90 to 110 mm* of mercury is frequently found in normal persons at all ages and is com patible with long life good health and a high degree of physical efficiency *In adults possessing blood pressures in this range essential hypertension is unlikely to develop Poor vasomotor control and systolic blood pressure of 90 to 110 mm do not neces sarily occur together* The symptoms of persistent fatigue dizziness and fainting

attacks are not due to persistent hypotension being frequently found in association with normal or elevated blood pressure These symptoms are frequently due to psychoneurosis

It is notable in psychoneurotic patients who have blood pressure of 90 to 110 mm that at such times as the anxiety or other underlying emotional disturbance is less evident not only are the general symptoms relieved but the blood pressure reading may be somewhat higher Conversely when the emotional disturbance again becomes prominent the blood pressure may gradually fall to the lower levels

Postural or orthostatic hypotension is characterized by the occurrence of faint ness or loss of consciousness on rising from the recumbent to the erect posture In this disorder the blood pressure may fall sharply from normal or elevated levels in the recumbent position to pressures of the order of 40 to 50 mm systolic and 25 to 40 mm diastolic in the erect position with loss of consciousness It is usually con sidered that postural hypotension is due to a failure of splanchnic vasoconstriction to offset the gravitational effects of the assumption of the erect posture

Orthostatic Tachycardia and Orthostatic Hypotension MacLean Allen and Magath have divided orthostatic tachycardia and orthostatic hypotension into two groups (1) *inconsistent orthostatic failure of venous return and* (2) *consistent* orthostatic failure of venous return

Inconsistent orthostatic failure of venous return is manifested by orthostatic weakness faintness and occasional syncope associated with orthostatic hypotension or tachycardia and an abnormal Flack reaction The Flack reaction is characterized by increased intrathoracic pressure the venous return fails to the extent that there are a marked decrease of the filling of the heart decreased cardiac output and sub sequent failure of the peripheral pulse Both signs and symptoms disappear in the recumbent position Hypotension if present is associated usually with compensatory tachycardia There may be only tachycardia in the erect posture associated with occasional drops of blood pressure or diminished pulse pressure Both signs and symptoms are usually much worse in the morning after the patient first gets out of bed and quickly improve after the subject continues to maintain the erect state At times during the day an abnormal Flack reaction may be the only indication that the patient has a potential defect of venous return

Usually no objective evidence of an organic dysfunction of the autonomic nerv ous system can be demonstrated The circulatory failure encountered in this incon sistent group appears to be associated with undue shifts of blood volumes and extracellular fluid gravitating to the lower extremities The increased heart rates low blood pressures and abnormal Flack reactions can be abolished by the assump tion of the recumbent position or by the application of tourniquets around the thighs It has been the experience of these writers that the loss of volume of circu lating blood through abdominal pooling appears to be a minor and relatively unim portant factor as compared with the loss related to the lower extremities

A normal man may exhibit at times the signs and symptoms of failure of venous return The margin between adequacy and incompetence is so narrow that when a man stands erect without moving he may be the subject of circulatory collapse Such failure may be precipitated by a variety of factors exposure to heat loss of blood poor tone of striated muscle psychic trauma or the maintenance of a re cumbent state for overlong periods

It is common to witness rapid heart rates and low blood pressures of patients who are allowed to stand for the first time after protracted rest in bed This postural instability is functional in that it is reversible If such a patient continues to stand erect for longer and longer periods each day vascular stability is regained The normal man can stand erect motionless for about one half hour (see Posture Chapter 6)

Orthostatic hypotensive tendencies and orthostatic tachycardia are observed

commonly during gestation Psychic syncope (fainting) seems to be largely postural and the cerebral ischemia can be prevented by the recumbent state Some patients have constitutional defects of venous return which may be termed neurocirculatory asthenia Occasionally the signs and symptoms of this disorder are orthostatic in origin and are characteristic of orthostatic failure of return of adequate amounts of venous blood to the heart

Caloric and rotational stimulation of the vestibules of normal man varieties of seasickness and airsickness the symptoms of which are postural undue exposure to heat abnormal loss of body fluid and primary shock with its nervous basis of pain and psychic factors all may demonstrate defects of venous return of varying degree which frequently play major roles in the total functional disability In these conditions the assumption of the erect posture may precipitate failure of venous return which is masked by the recumbent state

The pressure of hypertension is compatible with abnormalities of venous return Occasionally large drops in blood pressure of more than 100 mm of mercury are observed when a patient who has hypertension first stands erect after a period of rest in bed Such changes of blood pressure are as a rule transient lasting only a few seconds but during this period the Flack reaction may be markedly positive returning to normal when previous levels of blood pressure are established

Consistent orthostatic failure of venous return is manifested by severe orthostatic hypotension Objective evidences of disorders of the autonomic nervous system are common loss of sweating over large areas of the body surface associated with intolerance to heat frequent absence of compensatory reflex tachycardia and impotence Symptoms related to the deficit of venous return result from the erect posture or from application of a tourniquet to the femoral arteries of the legs The Flack reaction is invariably positive and frequently a deep inspiration will cause the peripheral pulse to disappear The signs and symptoms show classic exacerbation in the morning which is related to rest in bed Remissions are rare according to MacLean Allen and Magath

Hypertension (Hyperpiesis) The term hypertension refers to a state of elevated blood pressure both systolic and diastolic but frequently with only significant systolic increases in a patient who does or does not have obvious or determinable organic changes in the tissues The condition may be attended with associated symptoms or may be asymptomatic If symptoms are present they may be associated with the primary disease—for instance a glomerulonephritis—and not due to the hypertension

Incidence The frequency of hypertension increases steadily with age About one fifth of men between the ages of 20 and 29 years have readings of 140 mm systolic and 90 mm diastolic or higher The greater degrees of hypertension are relatively infrequent at these ages readings of 150 mm systolic and 100 mm diastolic or more occur in only 1 of every 20 cases

Hypertension of at least mild degree is common in middle aged and old persons Blood pressures of 140 mm systolic and 90 mm diastolic or higher are present in about 40 per cent of both men and women at ages 45 to 49 years and in 60 per cent at ages 60 to 64 years Readings of 150 mm systolic and 100 mm diastolic or higher are found in nearly 20 per cent of persons by the age of 45 years Thereafter the incidence rises steadily to more than 35 per cent in both sexes between the ages of 60 and 64 years Whatever definition of hypertension is used the condition is found more frequently in men than in women up to the age of 45 years After that age the incidence is generally higher in women

Transient Hypertension Levy Hillman Stroud and White made an analysis of the medical records of 22 741 officers of the United States Army in order to appraise the significance of *transient hypertension* Blood pressure readings higher than 150 mm of mercury systolic or 90 diastolic were co[illegible] abnormally high

The length of the observation period was from 1 year to more than 25 years 84 per cent were under observation from 5 to 19 years and 38 per cent from 15 to 19 years In 1 437 instances the duration of the observation period was 20 years or more

The frequency with which transient hypertension was first noted increased with age The curve of increase was smooth beginning with 5 9 per cent in the age group 25 to 29 years and reaching a plateau of 18 6 per cent at 50 to 54 years

At all ages sustained hypertension developed more frequently in those with previous transient hypertension than in those who never showed an elevation of blood pressure In both groups the rate increased with advancing years

The rate for disability retirement of an officer who had cardiovascular renal disease was consistently higher among those with transient hypertension than in those without at all ages from 35 to 60 years

The death rate with cardiovascular renal diseases was also higher in those with transient hypertension the figures rose in the older age groups

Hypertension and Endocrine Disorders Hypertension occurs as a symptom of *endocrine disease* and presumably results from hyperfunction of the thyroid and pituitary glands and from tumors of the medulla (paroxysmal hypertension) or cortex (Cushing s syndrome) of the suprarenal glands In hyperthyroidism there is frequently a rise in systolic pressure and pulse pressure which is thought to be due to the increased general metabolism In cases of hyperfunctioning tumors of the suprarenal medulla sometimes termed paragangliomas the elevation of blood pres sure usually but not always occurs in paroxysms There is evidence that the sudden marked rises in blood pressure are due to the release of epinephrine or closely allied norepinephrine from the tumor

Hypertension and Toxemia of Pregnancy In the toxemia of pregnancy (eclampsia) hypertension is a prominent feature A similar type of hypertension is present in a condition which Keith and Wagener have designated as acute vasospastic disease with hypertension

A primary glomerulonephritis or an essential hypertension may exist before the onset of pregnancy and may undergo exacerbation in the course of the pregnancy with resultant toxemic manifestation and these patients are best cared for if the primary renal or the primary vascular disease is recognized to exist along with the toxemia The patients in whom elevation of blood pressure occurs rapidly for the first time during pregnancy have a primary toxemia incident to pregnancy

Hofbauer has suggested that there may be a disturbed balance in certain hormonal regulatory mechanisms and homeostatic adjustments of normal gestation The various factors involved in the imbalance are to be seen in the hyperactivity of the pituitary the adrenal cortex and the accessory medulla like formation occurring in the cervical ganglia during gestation He has called attention particularly to the pronounced secretory activation of the chromaffin elements and their numerical increase in certain sympathetic ganglia situated lateral to the upper wall of the vagina and the cervix The cells of these ganglia rich in chromaffin elements display a close resemblance to the pheochromocytes of the adrenal medulla The structure appears to act during the gestation as an accessory adrenal medullary tissue functionally interrelated with the hypophysis the adrenals the ovaries and the placenta The histochemical appearance in the pregnant state of these pheochromocytes points to the increased formation of epinephrine norepine phrine or allied substances The enhanced chromaffin activity of these ganglia has a stimulating effect on the anterior pituitary adrenocortical system Certain physical factors in the second half of the gestation or during labor act mechanically to increase the release of epinephrine secreted by these cells into the circulation In preeclampsia Hofbauer points out pronounced eosinopenia sodium and chloride retention increased urinary 17 ketosteroids and uric acid excretion afford sensitive indicators of anterior pituitary adrenocortical stimulation with the activation of secretion of pituitary adreno corticotropic hormone most likely due to epinephrine

Hypertension and Arterial Degeneration Atherosclerotic degenerative changes of both the larger and smaller arteries may cause an elevation of the systolic blood pressure but there is no increase in the diastolic blood pressure in the former

Hypertension and Increased Intracranial Pressure Increased intracranial pressure may cause a rise in systolic blood pressure (160 170 mm Hg) but since such a mechanism is present for only a short time hypertension seems unlikely

Hypertension and Acute Vasospastic Disease Wagener observed and described visible vasospastic changes in the retinal arterioles in those who have eclampsia He and others have observed the same condition of the retinal arterioles in men and in nonpregnant women These patients have some of the symptoms of eclampsia They have an acute onset of symptoms with a rapidly increasing blood pressure from its normal or elevated state prior to symptoms These patients differ from those who have acute glomerulonephritis in that the evidence of renal damage is minimal or absent

Hypertension and Renal Disease Patients who have acute glomerulonephritis often have hypertension which begins early in the disease This hypertension may persist or it may disappear when the glomerulonephritis heals During this early period of hypertension and edema Keith observed that the fundi usually appear to be normal In an occasional instance however slight edema may be present about the disk margins and there may be present a few hemorrhages These mild retinal changes and hypertension may be secondary to the general toxemia which caused the nephritis including the fever

In those who have more severe acute nephritis improvement of the hypertension if present and general improvement as indicated by the disappearance of edema come slowly In these patients there usually develop chronic glomerulonephritis attended by a retinitis Some of them will have hypertension and some will not The retinitis develops as a result of edema anemia and angiospasm The patients who have both persistent moderate hypertension and generalized edema may have edema in the retina without fully developed retinitis

In those who have chronic glomerulonephritis with a minimal increase in blood pressure or perhaps no hypertension and in whom there is anemia of severe degree hemorrhages and cotton wool exudates may appear in the retina without angiospastic accompaniments Such a retinitis cannot be distinguished in general from that seen in anemia from any other cause However if the blood pressure is due to chronic nephritis the presence of persistent narrowing of the retinal arterioles may help to distinguish the retinitis from that which occurs in primary anemias

Angiospastic retinitis characterized by edema of the disks diffuse edema of the retina exudates of a cotton wool appearance hemorrhages star shaped macular lesions and visible sclerotic changes in the arterioles may be present in patients who have chronic glomerulonephritis with complicating or associated diffuse arteriolar disease and hypertension These retinal findings indicate that a diffuse arteriolar disease has preceded the onset of glomerulonephritis

These angiospastic and sclerotic lesions in the retinal arterioles indicate that thenceforward the vascular manifestations including the hypertension will assume the dominant role in the progress of the disease

Essential Hypertension The cause of essential hypertension has not been assigned The consensus is that abnormal physiologic processes which may give rise to hypertension comprise four factors (1) increased cardiac output (2) increased viscosity of the blood (3) increased total blood volume and (4) increased resistance in the peripheral circulation Increased cardiac output may be responsible for the hypertension found in some cases of aortic insufficiency and in patients with arteriovenous fistula However in most instances of essential hypertension the cardiac output is normal The viscosity and the total volume of the blood are in

creased in polycythemia vera whereas the blood pressure is normal or only slightly increased

An increased *peripheral resistance* that is a resistance in the arterioles or capil laries seems to be a prime factor in the production of the measurable arterial hyper tension This contention is based on histologic examination which reveals changes in the medial walls of peripheral arterioles throughout the body These changes consist of medial thickening which reduces the size of the lumina of the vessels A reduction in size of the lumina of these small vessels could give an increased resistance to the blood flow As soon as the postulation is made that there are present increased peripheral resistance and arteriolar changes sufficient to cause an increase in blood pressure there is no way to answer which came first the peripheral resistance or the arteriolar changes

Goldblatt s experiments on the production of renal ischemia in animals have revived interest in the old question of the relation of renal disease to diffuse arteriolar disease and hypertension The observation that hypertension will still develop in the ischemic kidney after the destruction of the nerve supply of the kidney indicates that there may be present an active hormone having its origin in the kidney and set free into the general circulation and that it is this substance which causes hypertension The nature of such a circulative substance and its mode of action are unknown Prior to Goldblatt s experi ments Tigerstedt and Bergman demonstrated that a simple saline extract of the fresh renal cortex of normal rabbits contained a pressor substance which they called renin This substance was not obtained from any tissues of the experimental animals other than renal tissues The site of action of renin appears to be in the peripheral arterioles and to be independent of the nervous system Despite these experimental findings on ani mals renin has not been demonstrated in the blood of a patient with hypertension

Interference with the blood supply of any organ other than the kidneys does not result in either temporary or permanent elevation of blood pressure Renal excretory insufficiency by itself is not enough according to Goldblatt for the development of hypertension In the early period of essential hypertension in man there is no renal insufficiency

Weitz observed the evidence of heredity in hypertension by measuring the blood pressure of brothers and sisters of hypertensive persons and found that the inci dence of elevated blood pressure was significantly greater among siblings of persons with hypertension than among siblings of persons without hypertension Ayman studied the blood pressure of every member of a family for three generations total ing 32 members more than 13 years of age One hundred per cent of the first gen eration 80 per cent of the second and 25 per cent of the third had elevated levels of the blood pressure on two or more readings Ayman in a later article reported on a direct study of the blood pressure height and weight of 1 524 members of 277 families Elevated systolic and diastolic blood pressure readings occurred in 148 of 780 members of the second generation aged from 14 to 39 years These 148 subjects had the same average age and sex incidence as the entire group of 780 children but they were 14⅓ pounds (6 5 kg) above the average weight compared to 4½ pounds (2 kg) above the average weight for the remainder of the group In families whose parents had absolutely normal blood pressures the incidence of elevated blood pressure in the children was only 3 1 per cent In families in which one parent had arteriolar hypertension the incidence of elevated readings in the children rose to 28 3 per cent In the families in which both parents had arteriolar hypertension the incidence of elevated readings in the children reached the level of 45 5 per cent Of 70 brothers and sisters of parents whose blood pressure was normal 37 3 per cent had elevated blood pressure readings whereas of 86 brothers and sisters of parents with arteriolar hypertension 65 3 per cent had elevated blood pressure readings This unusually high incidence of elevated blood pressure readings in the children brothers sisters and parents of subjects with arteriolar hypertension

as compared with similar relatives of subjects with normal blood pressure offers strong evidence for the existence of a hereditary factor in the etiology of hyper tension

Faulty diets have been associated with and assigned as a cause for hypertension in some patients The responsible factors in or absent from the diet have not been determined

The group predisposed toward hypertension on a hereditary basis must fre quently acquire faulty dietary habits as habits of food selection are handed down through families with little change from one generation to the next The effect of diet and overweight is also well demonstrated by the benefit incident to correction of these factors in patients in whom hypertension has developed

Increased blood pressure is produced by *anger and fear* Stimulation of the sympathetic nervous system in anger and fear by an increased amount of norepine phrine and epinephrine in the blood stream causes rise in blood pressure The repression of chronic fear or chronic rage *according to the psychiatrist eventually* manifests itself in the circulatory system by the production of a continuous state of increased tension through the mechanism of sympathetic nerves and norepine phrine and epinephrine induced generalized vasoconstriction

Long continued increased tension eventually produces degenerative changes in the intima and thickening of the walls of the arterioles Although these changes fre quently occur as one of the degenerative changes of increased age they are appar ently greatly speeded by hypertensive states Changes are generalized and include the afferent renal vessels

It has been suggested that hypertension is due to abnormal development of arteriovenous fistulas such as normally exist in the tips of the fingers and toes and in certain parts of the kidneys There is little evidence to support this hypothesis

Hines and Brown interested in the vasomotor factors in hypertension devised a standard test for measuring the maximal vasomotor response when the hand and arm were immersed in cold water Individuals who have essential hypertension show a much greater increase in pressures with the cold water test than those who do not have essential hypertension Among patients who had early or preorganic hyper tension the average increase of blood pressure was 34 mm of mercury systolic and 25 mm diastolic Among the patients who had an organic stage of essential hyper tension the average increase of blood pressure was 47 mm of mercury systolic and 34 mm diastolic

As a result of a study of 190 members of 15 family groups 6 without and 9 with definite evidence or history of hypertension Hines and Brown expressed the opinion that the vasomotor reaction to cold follows an inherited pattern and that the exces sive or hypertensive type of reaction occurs in the families in which there is a hypertensive diathesis They were led to believe that the abnormality of essential hypertension is an excessive response in the blood pressure to intrinsic and extrinsic stimulation This hyperreactive vasomotor mechanism may be an important factor in the production of arteriolar hypertrophy and in the subsequent development of the organic stages of the disease

Hines and Brown found that the systolic form of hypertension seen in cases of senile arteriosclerosis and in cases of glomerulonephritis gave definitely less response to the cold pressor test than did the pre existent and existent stages of essential hypertension

For a long time comments have been made in regard to the temperament which was apparent early in the life of hypertensive patients These children are tense and more conscientious than the average child of their age On examination of such a child there is present evidence indicating vasomotor weakness such as frequent epistaxis excessive menstruation cold sweaty and cyanotic hands flu hing blink ing of the eyes and other evidences of a high strung temperament

PATHOLOGY Barker has summarized the pathologic changes in hypertension as consisting primarily of the development of a spastic thickening of the walls of the arterioles throughout the body which gradually becomes organic The secondary manifestations are myocardial weakness and failure as the result of increased myocardial strain and the premature development of degenerative changes in the large arteries also as the result of strain The degenerative changes in the larger arteries occasionally result in rupture more commonly in thrombosis and either of these events may be serious or fatal if the vascular accident occurs in a vital organ such as the heart or brain Serious lesions of the kidneys may occur as the results of chronic ischemia in hypertensive patients When death occurs it is most commonly the result of myocardial failure second most commonly the result of cerebral vascular accidents and third most commonly the result of renal insufficiency

CLASSIFICATION OF ESSENTIAL HYPERTENSION The most widely known classification of essential hypertension is Volhard s red and pale hypertension Keith and Wagener admit the merits of Volhard s basic conception of these two conditions but they are convinced that it is more appropriate to classify hypertension on the evidence of the differences in the arteriolar changes In support of the soundness of their classification which is based on changes in the retinas they took biopsy specimens from striated muscle and found a close correlation between arteriolar changes in the muscle and those revealed on ophthalmoscopic examination Muscles were selected for the purposes of biopsy because this tissue represents one of the large bulky systems of the body

Their histologic study of biopsied muscle disclosed that in some cases the arterioles appeared to be normal whereas in many others the walls of the arterioles were thickened the ratio of the diameter of the lumen to the thickness of the wall was definitely decreased In many instances this change was uniform for all the arterioles seen In some it was confined to the smallest arterioles and in others there were marked variations and localized changes only Other changes consisted of increased prominence and tortuosity of the arterioles and an increase in the number and size of medial and intimal nuclei Occasionally there were organized thrombi extensive intimal proliferation complete occlusion of the lumen by simple hypertrophy of the media and perivascular collections of lymphocytes and fibroblasts Medial necrosis was found in the arterioles of voluntary muscle as well as in those of the kidneys and other organs in malignant hypertension None of these changes were found in the histologic sections of muscles of persons with normal blood pressure Similar findings to those found in the arterioles of striated muscle were demonstrated in the arterioles of the retina

Keith and Wagener s classification of essential hypertension is based on the foregoing studies which emphasize the widespread dissemination of the arteriolar changes A view of the pathologic processes present in the arterioles in a particular patient who has hypertension can be had by ophthalmoscopic examination a method of examination available to any patient Therefore the findings on general and on ophthalmoscopic examinations determine to which of the four groups of hypertension the patient is assigned The division of hypertension into four groups by these observers is based on the following criteria

Diffuse Arteriolar Disease with Hypertension Group 1 These patients are usually between 20 and 55 years of age Members of this group constitute the great majority of those who have hypertension and they often have good health for years

The evidence of the hypertension often is first found at the time of routine examination Prior to such an examination there have been no symptoms There are no symptoms in the stoic individuals after they know the blood pressure is increased It is different in the psychoneurotic individuals who often become hypertensive hypochondriacs

On examination there may or may not be obesity The blood pressure is 150 to 175 mm of mercury systolic 90 to 95 mm of mercury diastolic Often the diastolic

pressure is normal The heart is not enlarged or seemingly abnormal in any way When the radial artery is made pulseless at the wrist by compression at the elbow the pulseless artery may or may not be palpated If it should be palpable it imparts the sensation of a soft collapsible tube to the palpating finger

Twenty four hour studies of blood pressure demonstrate the decidedly beneficial effect of rest on the hypertension During rest and sleep the blood pressure is often within normal limits and the retinal changes are usually minimal and consist of mild narrowing or mild sclerosis of the arterioles Spasm of the arterioles is not present

The changes in the arterioles of the muscles likewise are minimal Examination of the urine reveals no abnormality or at most the urine contains albumin grade 1 on a basis of 1 to 4 in which 1 indicates the least amount of albumin and 4 the greatest amount More than albumin grade 1 arouses the suspicion of a previous nephritis The urinary sediment is negative

Diffuse Arteriolar Disease with Hypertension Group 2 These patients like those of group 1 vary in age from about 20 to 55 or more years They may be obese

If these patients are ill it is not from their hypertension However the blood pressure is distinctly elevated 160 to 185 or more systolic and 90 to 100 or more diastolic Some of the patients may have headaches Nocturia is often present particularly in the aging patients The heart may or may not be enlarged Obliteration of the radial pulse by pressure at the elbow reveals a definitely palpable thickened radial artery at the wrist Often flexion of the elbow will reveal that the arteries of the arm have lost a considerable part of their elasticity and do not contract with the flexed arm They seem to be too long and are observed to be tortuous tubes pulsating in two planes

There is usually a response to rest by the blood pressure perhaps a return to normal values will be revealed while the patient is asleep

Patients of this group have continuously a higher blood pressure than normal and more distinct changes in their retinal arterioles The arteriolar sclerosis is often graded 2 to 3 on a basis of 1 to 4 There is no or at most minimal angiospasm

Histologic studies of the muscle arterioles disclose a more uniform and more marked narrowing than is evident in the arterioles of group 1 In addition to narrowing of the lumen of the vessel there may be thrombosis or occlusion of arterioles

The urine usually contains albumin grade 1 to 2 The urinary sediment often contains hyaline and granular casts but rarely microscopic blood

As these patients pass into the aged groups of individuals atherosclerosis is often superimposed and any one of the cerebral cardiac or renal accidents incident to this disease may occur The impression is gained that a person who has arteriosclerosis with hypertension is more susceptible to atherosclerosis than one who does not have arteriosclerosis

The clinical impression is gained that the patients of group 2 have a progressive disease The hypertension of patients in group 2 more nearly corresponds to Volhard s designation of red hypertension

Diffuse Arteriolar Disease with Hypertension Group 3 These patients are between 20 and 55 years of age and men are more commonly affected than women

Generally there are headaches feelings of fullness in the head failures of vision nosebleed dizziness and unsteadiness of gait Often there is exertional dyspnea cough or other symptoms of left sided heart failure Nocturia and polyuria are common All of these symptoms may increase rapidly then remain stationary for a time and then regress but more often they tend to progress and increase in intensity

On examination these patients often have a general pallor the so-called pale hypertensives of Volhard There is often evidence of loss of weight The blood pressure is high 200 to 300 mm of mercury systolic and 100 to 130 mm or more diastolic are common The radial arteries however may not reveal much arterio-

sclerosis Often the sclerosis is not more than grade 2 on a basis of 1 to 4 The pulse rate is often increased The heart is enlarged and the aortic second sound is accentuated Basal aortic systolic murmurs are common Edema of the ankles may be present after the patient has been on the feet for an hour or two The urine contains albumin and often the presence of blood and a few casts may be revealed by microscopic examination The concentration of urea nitrogen in the blood may be increased depending on the degree of renal involvement More often there is present an inability of the kidneys to concentrate urine The specific gravity of the urine has a limited range of variability (1 010 to 1 015)

An important criterion is the presence of angiospastic retinitis together with definite sclerotic changes in the arterioles but no edema of the disks In those who have had a remission from the progress of the disease there may be evidence in the retina of previously active retinitis indicated by the obliteration of small arterioles In such cases small arterioles of the retina undergo thrombosis and a similar process may have occurred in the skeletal muscles

On examination of the arterioles of the skeletal muscles the lumina of the arterioles are decreased in size definite cellular lesions and thrombosis are often present

Diffuse Arteriolar Disease with Hypertension Group 4 (Malignant Hypertension) This disease affects men more frequently than women The onset often occurs between the ages of 20 and 55 years Malignant hypertension has been reported however in older individuals and in children

These patients are nervous tense and weak and complain of visual disturbances or more likely faulty vision or definite failures of vision Headaches are often intense and may approach in severity those caused from intracranial hypertension

Dyspnea palpitation and some enlargement of the heart are often present The radial artery may be sclerosed grade 2 or 3 on a basis of 1 to 4 Blood pressures range from 200 systolic upward measured in millimeters of mercury and the diastolic may be 130 to 140

The urine contains albumin and often blood In an occasional instance there may be profuse renal hemorrhages which are sufficient for blood clots to form in the ureters and cause acute colic These clots are usually expelled spontaneously

The important retinal alterations are the marked spastic and organic narrowing of the arterioles with diffuse retinitis and edema of the disks The edema of the disks if accompanied by the severe headaches may require differentiation from increased intracranial hypertension

The arterioles of muscle obtained by biopsy reveal great narrowing of the lumina cellular lesions and often thrombosis or occlusion

The prognosis is serious only one of Wagener and Keith s patients lived as long as 40 months Seventy nine per cent were dead within a year However one patient was alive 11 years after the first observation

Not in all cases of group 4 is the course so rapid Some patients even have periods with subsidence of the retinitis and with clinical symptoms which suggest remission Such cases belonging to group 3 were considered previously Here again in group 4 spasm of the arterioles may determine whether the condition is progressive

Differentiation of Groups The numbers 1 to 4 were employed by Wagener and Keith to avoid confusing terminologies The numbers 1 to 4 do not imply a progression from 1 to 4 in all instances of hypertension The number used designates or classifies the group of hypertension to which the patient belongs at the time of the first examination In reality a patient belonging to group 1 may suddenly become a member of group 4 or a patient belonging to group 3 may become a member of group 1 and in very rare instances one who has hypertension group 4 may have a remission in the retinal findings and be classified as belonging to group 3

Clinical differentiation of the four groups of essential hypertension one group from another may not be accomplished without difficulty Patients of groups 1 and 2 have good retinal cerebral cardiac and renal function whether the hypertension be labile or high with a tendency toward fixation In those belonging to group 3 there is evidence of dysfunction of one or several of the organs belonging to the arterial system while in group 4 these functional disturbances become more effective and final failure may occur because of a simultaneous serious interference in the blood supply to the retina brain heart and kidneys Difficulties of differentiation of one group from another arise when in a given case the disease progresses rapidly so that the patient passes from group 1 to group 4 in a short period

Examination of the ocular fundi is of considerable aid in determining the group in which an individual patient with hypertension belongs *For practical clinical purposes* those who have only purely organic changes in the arterioles of the retina belong to group 1 or group 2 while patients who show definite evidence of angiospasm belong in group 3 or group 4

Thrombosis of the retinal veins is considered to arise either on the basis of organic changes alone or as the result of an infectious process (phlebitis) superimposed on a mild organic lesion Patients who have retinal venous thrombosis usually belong in group 2 although the other clinical and laboratory findings at times may justify their classification with hypertension of group 3 Cases of retinitis of arteriosclerosis usually can be placed in group 2 unless the retinitis can be shown definitely to be the residuum of a previously angiospastic type of retinitis

Patients who have retinitis of angiospastic type characterized by edema cotton wool appearing exudates and hemorrhages in the retina superimposed on a combination of sclerotic and spastic lesions in the arterioles belong to hypertension group 3 or group 4 Patients who have cotton wool appearing exudates and hemorrhagic areas in the retina but whose optic disks are not edematous usually belong to group 3 The most characteristic feature of the disease in cases of hypertension of group 4 is measurable edema of the optic disks which usually occurs of course in association with the other features of widespread angiospastic retinitis and in intracranial hypertension

Acute angiospastic retinitis is a part of vasospastic disease which is often present in the toxemias of pregnancy and active severe arteriolar sclerosis The ophthalmoscopic findings in these conditions vary from those of localized areas of edema of the retina with cotton wool appearing exudates and hemorrhages to that of retinitis with diffuse edema of the retina In both acute angiospastic disease and toxemia of pregnancy the retinitis may completely disappear under favorable circumstances leaving as residuals varying grades of sclerosis of the retinal arterioles depending on the severity and persistence of the underlying angiospasm The features of an angiospastic disease which may clear up depend on the presence of angiospasm generalized and localized irregular narrowing of the arterioles and absence of organic changes in the vessel walls such as visible thickening of the wall exaggerated reflexes and arteriovenous compression In an angiospastic disease when organic changes are developing in the walls of the arterioles differentiation of group 3 or group 4 may be difficult or impossible without the presence of edema of the disks as regularly occurs in group 4

Wagener and Keith have expressed the belief that in the beginning of a toxemia of pregnancy the differentiation between retinitis of hypertension and albuminuric retinitis (nephritis) cannot be made except by the time element since in most patients who show primarily a few cotton wool patches and hemorrhages the characteristic appearance of albuminuric retinitis will later develop unless the pregnancy is terminated spontaneously or otherwise In the pregnant woman as in the nonpregnant woman the albuminuric type of retinitis is more often the result of diffuse arteriolar disease than it is of primary glomerulonephritis

Keith and Wagener have presented evidence to support the contention that the development of anoxemia in the retina is demonstrated by the appearance of retinitis and that the retinal tissue lysins resulting from the anoxemia secondarily cause organic changes in the vessel walls However they have observed that permanent organic changes at times may take place in the arteriolar walls after persistent spasm without the development of retinitis

Prognosis Many of those who belong to groups 1 and 2 live their normal life expectancy However these patients often pass on to middle or old age and then atherosclerosis develops and thus heart failure renal failure or strokes ensue

The prognosis for patients in group 3 and especially for patients in group 4 is grave A death rate within one year of one third of those in group 3 and about two thirds of those in group 4 is in distinct contrast to that of 1 of every 10 in groups 1 and 2

The prognosis in a group of patients who have hypertension from various causes has been forecast by Frant and Groen who reported a follow up of 418 patients with hypertension (blood pressures above 155 systolic and 100 diastolic) who were re examined after a period of 8 to 9 years The death rate for men with essential hypertension exceeded the normally expected death rate in the same age groups by 102 per cent for the women this figure was 91 per cent Chronic nephritis increased the death rate in men by 587 per cent and in women by 150 per cent Hypertension related to toxemia of pregnancy carried a mortality of 155 per cent in excess of the normal The total excess mortality for patients with hypertension of all types was 233 per cent for the men and 201 per cent for the women The mortality increased with increase of both systolic and diastolic blood pressure

The condition of the fundus of the eye is a better guide for the prognosis of hypertension than the increase in blood pressure In the cases studied by Frant and Groen heart disease albuminuria and diabetes appeared to reduce the expectation of life even more for women than for men Hypertension accompanied with obesity appeared to be prognostically more favorable than the same hypertension in patients whose weight was normal or low Hypertension in young subjects carries a relatively shorter life expectancy than high blood pressure in old age Heart disease is first among the causes of death of patients with hypertension it accounts for 40 9 per cent next comes carcinoma (16 4 per cent) followed by uremia (14 9 per cent) and apoplexy (8 6 per cent) Apoplexy as cause of death was almost five times as common among women (14 2 per cent) as among men (3 1 per cent) The cause of death could not be ascertained for 9 4 per cent

The prognosis seems to be better since the advent of new drugs

Arteriosclerosis and Atherosclerosis With and Without Hypertension Hypertension with an increase in diastolic pressure is probably not present without disease of the smaller arteries or arterioles Indeed on general examination there may be evidence present of disease of the larger and smaller arteries without an accompanying hypertension Determination of the basal blood pressure reveals a pressure within the limits of normal for the age group to which the patient belongs However these observations if made in the presence of manifest cardiac or renal disease may be diagnostically misleading for such patients may have had a very low normal basal pressure prior to the development of manifestations of cardiac or renal disease Such observations require definitions of arteriosclerosis arteriolar sclerosis and atherosclerosis

Definitions of arteriosclerosis arteriolar sclerosis and atherosclerosis are difficult for these terms have been variously employed to mean different forms and degrees of vascular disease as well as the same vascular disease

The term arteriosclerosis (hardening of the arteries) in its broad use refers to degenerative vascular changes which are not due to syphilis rheumatic fever or specific infectious diseases and the term does not include the conditions known as thromboangii

tis obliterans periarteritis nodosa and lupus erythematosus Arteriosclerosis designates a condition marked by loss of elasticity thickening and hardening of the arteries Appropriate adjectives designate the region of the body most profoundly affected for instance cerebral coronary or in the case of widespread disease general arteriosclerosis A special form in medium sized and smaller sized arteries characterized by a primary necrosis and calcification of the medial coat is termed Monckeberg s sclerosis

As the term arteriolar sclerosis commonly is used it designates changes in the *smaller arteries* and *arterioles* which are characterized by endothelial hyperplasia thickening and hyalinization of the intima and hypertrophy of the media of the arterioles of the kidneys and other viscera and which are more uniformly distributed in the arterioles than is either arteriosclerosis or atherosclerosis in the larger arteries Arteriolar sclerosis is always associated with hypertension These arteriolar changes are of the type of vascular disease responsible for essential hypertension (groups 1 to 4) of Wagener and Keith

Atherosclerosis affects the *large* and *medium sized arteries* such as the aorta and the coronary and cerebral vessels the arteries to the arms and legs and the larger peripheral arteries The condition is characterized by intimal thickening due to lipoid atheroma fibrosis calcification necrosis and hemorrhage This is the type of arteriosclerosis which involves the coronary and cerebral arteries and is often responsible for arteriosclerotic heart disease and cerebral apoplexy in those who do not have hypertension

Atherosclerosis is common after the age of 40 years There is an increasing frequency with subsequent decades

Heart disease renal disease and cerebral disease due to arteriosclerosis and atherosclerosis are much more frequent among men than among women

Hypertension is present in about one half of all the men and in about three fourths of all the women who have these degenerative forms of heart disease Arteriosclerotic heart disease in the absence of hypertension suggests that hypertension is an aggravating factor and not the cause

Arteriosclerotic heart disease develops with unusually high frequency among diabetic patients and those who have familial xanthomatosis with hypercholesterolemia myxedema or chronic glomerulonephritis and obesity

The theories of the etiology of arteriosclerosis and atherosclerosis are concerned primarily with whether or not the disease is a process of aging or with conditions incident thereto such as metabolic disorders inherited or acquired

The life span of man no doubt does change according to various and unknown conditions arising within his own body or as the result of environmental conditions It would be well to wait before conclusions are drawn concerning the increasing span of human life as well as the purported increase in the incidence of arteriosclerosis and hypertension to see whether these might not be just passing phases in the evolution of the species To gloat over any particular achievements of this generation over the immediately former generations might prove to be premature

In a similar vein the tempo of modern industrialized civilization is indicted as the cause of hypertension and hardening of the arteries A comparison of the tempo and the emotional tensions of men who lived 25 000 or more years ago is not available If such were available might it not be just possible that these men and women had many tensions unknown to us now? Did they not have arteriosclerosis and hypertension often resulting in sudden death? One can be sure that if they did have angina pectoris or strokes they were not accorded protection in bed for prolonged periods of time The prolongation of such a life by bed rest alone is hardly worthy of praise as a medical accomplishment

Transient and occasionally permanent hypertension may follow repeated powerful emotional experiences involving fear anxiety and hostility A characteristic hypertensive personality and a distinctive psychiatric pattern have been described

Hypertension is slightly commoner in women than in men Hypertensive (and arteriosclerotic) heart disease is much more frequent in men

GENETICS AND ATHEROSCLEROSIS Adlersberg Parets and Boas have recorded 35 families with xanthoma and the relationship of this condition to hypercholesterolemia coronary artery disease and related stigmas It appeared to these authors that a common factor for some patients with coronary atherosclerosis may be a hereditary disturbance of cholesterol metabolism manifested by elevated serum cholesterol Familial xanthomatosis is the severest form of the inherited disturbance Xanthomatous lesions develop only in patients who carry two abnormal genes for cholesterol that is they are homozygotes Atherosclerosis is frequent in such persons These authors believe as others do that some patients with uncomplicated coronary artery disease are probably affected with a mild form of disturbed lipid metabolism They carry one abnormal gene for cholesterol that is they are heterozygotes Derangement of cholesterol metabolism helps explain the familial incidence of coronary artery disease and also accounts in part for its development in many persons less than 50 years of age

METABOLISM The only constant factor in all the metabolic theories of arteriosclerosis is that cholesterol and other lipids play a common role in the production of the disease It is thought that the initial disturbance in atherosclerosis is one of solution or dispersion of cholesterol in the blood due either to hypercholesterolemia or to alteration in other substances in the blood which diminish the solubility of cholesterol

There are large amounts of cholesterol esters and free cholesterol in atheromatous lesions The quantity present exceeds that which could be formed by local degeneration The concentrations of lipids in these lesions correspond to their concentration in the plasma

Gofman and associates have pointed out that it is not the total plasma cholesterol which correlates with vascular sclerosis but rather the presence and quantity of an abnormal cholesterol containing lipoprotein

Since dietary cholesterol occurs in foods of animal origin with particularly desirable nutritional value (for example meat eggs and milk) it seems unwise on the bases of the present evidence to condemn use of these foods if there is risk of sacrificing the foundations of sound nutrition This is particularly significant and in accord with the observations of Keys and associates who observed that the serum cholesterol level of normal men is not significantly related to differences in the habitual cholesterol intake over a range of something like 250 to 800 mg per day

Premature atherosclerosis occurs in diabetes However the specific metabolic or endocrine factor in diabetes which predisposes to atherosclerosis is uncertain

OBESITY AND ATHEROSCLEROSIS Faber and Lund studied 408 aortas from necropsy material in hospitals and the medicolegal institute of Copenhagen Two hundred and ninety five of the 408 patients 240 men and 168 women from whom the aortas were obtained had been of normal weight and 113 had been obese Two hundred and eighteen of normal weight and 41 obese persons had had normal blood pressure 77 of normal weight and 72 obese persons had had hypertension The great increase in the degree of sclerosis with rising age makes it necessary to eliminate the age factor from the evaluation of the sclerosis The logarithm of the cholesterol and calcium content of the vessel wall and the dry weight of the tissue rises rectilinearly with age To eliminate the factor of age the formula for this line had been calculated for these three factors Distribution curves for the dry weight the total cholesterol content and the calcium content of the intima and media of the aorta around the calculated normal line for the four groups of persons studied were constructed on the basis of the deviations from this line Comparison of these distribution curves showed that hypertension gives a rise above what should be expected according to age Obesity itself has no effect on any of the factors studied when the presence of hypertension is taken into account

MANIFESTATIONS The manifestations of arteriosclerosis or atherosclerosis de

pend on the individual arteries affected and their distribution to various members organs or systems of the body These symptoms are discussed in association with the organs or systems affected

Symptoms arise when there is interference with the blood supply to the part caused by occlusion partial or complete or rupture of the artery Associated or unassociated with partial or complete occlusion or rupture of an artery may be disturbances in conduction and rhythm of the heart in cerebral functions in digestive functions and in locomotion attributable to atherosclerosis

On examination either arteriosclerosis or atherosclerosis may be found without the demonstrable presence of the other Arteriosclerosis often will be accompanied by hypertension Arteriolar sclerosis will always be accompanied by hypertension These conditions are diagnosed by ophthalmoscopic examination

Examination when the arm is flexed at the elbow reveals a visible tortuous brachial artery which pulsates in two planes for the artery has lost its elasticity Often palpation of such an artery reveals small hard irregular plaques The pulsations in such arteries especially when those of the legs are involved may reveal decreased or absent pulses

The roentgenograms often reveal the presence of radiopaque material along the course of the larger arteries in all parts of the body especially the aorta and the pelvic vessels

The diagnosis of arteriosclerosis and atherosclerosis depends on the symptoms and physical signs resulting from the diminishing or the discontinuing of blood flow to the affected part

The peripheral arteries can be palpated and often calcification in their walls can be demonstrated by roentgenologic examination

The presence of arteriosclerosis in a youth suggests the possibility that diabetes mellitus is present

Angina Pectoris The immediate cause of angina pectoris is insufficiency of the coronary circulation

The causes of insufficiency of the coronary circulation as enumerated by White are (1) diseases of the coronary arteries due to atherosclerosis infection thrombosis or embolism (2) the coronary arteries may be blocked at their mouths partially or completely by swelling and inflammatory reaction of the aortic wall in syphilitic aortitis or by vegetations on the aortic intima or ex ending up from the aortic valve cusps in bacterial endocarditis There may be (3) marked aortic regurgitation with very low diastolic blood pressure it is an ample diastolic blood pressure that normally maintains an efficient coronary circulation for during systole the coronary arteries are compressed There may be (4) marked aortic stenosis with low pulse pressure and limited output per beat which doubtless with the added effect of suction by the forceful narrow jet of blood directed up the aorta with each heart beat cuts down the coronary circulation There may be (5) severe anemia pernicious or secondary with insufficient oxygen in the blood carried to an active myocardium even though the coronary arteries themselves are fairly normal There may be (6) too great a demand on the heart muscle as in excessive exertion or thyrotoxicosis so that the coronary circulation though adequate at rest is unable to maintain a sufficient blood flow to the myocardium under these circumstances There may be (7) spasm of the coronary arteries lasting a few minutes at a time in patients who have relatively normal vessels

Hypotheses of Pain in the Heart There are two often quoted hypotheses of the origin of cardiac pain ischemia of heart muscles and tension on the coronary arteries The following paragraphs summarize the observations of Blumgart on the origin of cardiac pain due to ischemia

Ischemia In the syndromes of angina pectoris coronary failure and acute myocardial infarction cardiac pain is an expression of a relative disproportion between the demands of the myocardium for blood and the decreased supply it receives through the coronary arteries The changes in the myocardium resulting from this disproportion depend on the duration of the ischemia If the ischemia is brief the j

angina pectoris and no permanent myocardial damage may occur If the ischemia is more prolonged the pain is more severe and prolonged If the duration of complete obstruction is longer than 15 minutes irreversible changes that is myocardial necrosis are produced notwithstanding re establishment of normal blood supply When the blood supply has been interrupted for 40 to 45 minutes widespread myocardial infarction is witnessed during the succeeding days even though blood flow through the coronary arteries is not obstructed

Cardiac pain is precipitated by effort emotion and other states which increase the work of the heart and is relieved by rest and other measures which lessen the work of the heart Cardiac pain may be induced by breathing low concentrations of oxygen All of these facts support the concept that cardiac pain is due to ischemia The structural changes in the coronary arteries and in the myocardium must not be considered the exclusive cause of cardiac pain but rather as constituting the stage on which various factors may operate Thus vasoconstriction or absence of vasodilatation effort anemia tachycardia or shock with its lowered blood pressure may act as precipitating agents in the production of pain in a heart whose circulation is already compromised by arterial obstruction

Tension According to Gorham the arguments for the tension hypothesis of cardiac pain have been advanced and supported by such evidence as the occurrence of coronary occlusion without pain Pain is more frequent in the presence of acute fresh thrombosis and infarction and is less frequently encountered in old fibrous infarcts without actual thrombosis

Pain is known to arise from blood vessels owing to tension on the walls of arteries in instances of embolism of the peripheral arteries in dissecting aneurysm and in migraine

Tension is the commonest cause of pain as noted in inflammatory lesions and in the colics produced by smooth muscle contraction in hollow organs

Pain may be produced in the human heart affected with coronary sclerosis and the anginal syndrome by the injection of epinephrine

Angina pectoris occurs predominantly in men The disease occurs in women usually in association with diabetes or hypertension The average age of onset is 56 years for men and 58 years for women

There are no convincing data to prove that angina pectoris affects particularly those whose work subjects them to continuous mental tension Angina pectoris occurs in individuals of all races and all social and economic strata

The genealogic history often discloses that the patient s progenitors in one or more lines of the family suffered from angina pectoris Of the affected line or lines of the family the members who have angina pectoris are usually short and stocky overweight short necked with barrel shaped thorax

These patients are aggressive ambitious serious minded persons who possess deep emotional reactions

PATHOLOGY The pathologic changes are varied They are those of any disease or condition which causes a narrowing and occlusion of the lumina of the coronary arteries Diseases and states causing coronary narrowing are atherosclerosis syphilitic aortitis aortic valvular disease reduction in aortic blood pressure pulmonary embolism shock states reduction in oxygen content of the blood as in anemia and arterial anoxemia as caused by pulmonary or congenital heart disease low atmospheric oxygen tension and drugs

SYMPTOMS Angina pectoris is characterized by paroxysmal attacks of pain or oppression situated retrosternally and extending commonly to the precordium and left pectoral girdle usually to the arm

The pain in angina pectoris is as a rule dull and throbbing is produced by effort and is relieved immediately by rest or nitrites The pain of acute myocardial infarction is distinguished from that of angina pectoris by being a more prolonged and usually more severe thoracic pain

The occurrence of the pain or pressure with effort is the essential feature of the syndrome The pain of angina pectoris rarely occurs while the patient is at rest

In such a patient if the pain or oppression cannot be reproduced by exertion or sexual intercourse the diagnosis of angina pectoris is dubious

The pain during a paroxysm of angina pectoris is usually situated at or behind the middle or upper third of the sternum centered at the level of the third or fourth rib Occasionally the pain is more extensive and covers most of the precordium A pain that is situated at or near the cardiac apex or to the left of the cardiac region is not due to angina pectoris

The pain of angina pectoris when it is severe tends to extend to the arm neck and occasionally to the jaw In its most precise form the pain extends across the precordium to the left shoulder and upper medial side of the arm and may extend as far as the elbow the wrist or the fingers It may affect the wrist and fingers without involving any other portion of the extremity Sometimes the pain starts in the left wrist arm or shoulder and travels toward the sternum It may commence in the sternal region and pass directly toward the left scapular region

The more severe pain of angina pectoris may be described as constrictive pressing choking or expanding There may be a sense of a vise around the thorax Often the pain resembles that of gaseous distention and the patient tries to belch and takes baking soda in attempts to obtain relief The pain may or may not compel the patient to stop activity and to rest

A common characteristic reaction of the patient to the pain is the fear of immediate impending disaster termed angor animi

Duration of the attack of angina pectoris which occurs during physical activity is less than 5 minutes The attacks which occur while the patient is at rest last longer often from 5 to 15 minutes or up to a half hour When severe pain lasts for more than a half hour an acute coronary occlusion is usually present

The frequency of the attacks of angina pectoris varies the intervals depending to some extent on avoidance of the factors which excite the attacks Attacks may come each day without apparent cause or after performance of the essential physiologic functions

Walking especially against cold wind or walking rapidly is the commonest form of bodily exertion producing the pain of angina pectoris Under favorable walking conditions on level ground one of these patients may walk a mile or more without discomfort Some patients have pain when they walk for only a short way during the early morning hours and then are able to perform rather heavy manual work the rest of the day

After a meal less effort is required to provoke an attack than when the stomach is empty Straining at stool is sometimes responsible for an attack Any or all emotional upsets such as heated arguments the excitement of card playing grief and anger and a variety of petty annoyances may induce an attack Not infrequently cardiac pain occurs during sexual intercourse

Excessive insulin dosage may induce attacks of angina pectoris

Examination There are no abnormal physical signs in patients who have uncomplicated angina pectoris

The blood pressure in about half of those who have angina pectoris is within the normal range The rest have hypertension

Ordinarily electrocardiographic studies are made while the patient is free from pain then the tracings are often entirely normal There may be abnormalities however of the T and Q waves or of the QRS complexes

Tests of Coronary Reserve and Coronary Insufficiency There is a range in which increases and decreases of the amount of the coronary blood flow to the myocardium occur without the production of pain This range represents the coronary reserve In those who have an impaired range of adjustment of the coronary circulation the coronary reserve is diminished Attempts have been made to establish for diagnostic purposes tests or methods which measure the degree of

insufficiency of the coronary circulation These tests are (1) exercise in graduated amounts and (2) the anoxemia test Usually these tests are not required and in a patient who has definite coronary disease performance of any test is undesirable (see Tests of Cardiac Function page 704)

The patient who has angina pectoris may scarcely be inconvenienced or his activity may be involuntarily so restricted that he is a chronic cardiac invalid Most patients for at least a few years after the initial episode remain capable of earning their livelihood at their usual occupation as they become accustomed to the range of bodily activities which can be performed without having pain

DIAGNOSIS The essential diagnostic features of angina pectoris are (1) the paroxysmal occurrence of the pain (2) its brief duration (3) its characteristic location radiation and quality and (4) its precipitation by effort and relief by rest or nitrites

The diagnosis of angina pectoris should be made with reservation in a woman unless there is evidence of an underlying disease capable of producing coronary insufficiency

In a series of cases Parker Dry Willius and Gage observed that the average age at onset of the anginal syndrome was slightly earlier for men than for women While there was wide variation in age of the patients at the onset of symptoms 88 per cent of the patients were from 40 to 69 years of age The ratio of men to women was 4 3 1

The highest mortality rate occurs in the first year following the establishment of the diagnosis of coronary arterial disease with angina pectoris Thereafter the yearly mortality rate is less than it was in the preceding year The survival rates of women having the anginal syndrome are greater than those of men

When corrected for deaths not due to angina pectoris the five year survival rate of patients 30 to 39 years of age with the anginal syndrome is definitely shorter than when the disease manifests itself later in life

Such associated conditions as cardiac hypertrophy well defined hypertension previous cardiac infarction congestive heart failure and significant electrocardiographic abnormalities are clearly related to a higher mortality rate and lower survival rates

Acute Coronary Occlusion and Myocardial Infarction An acute thrombotic occlusion of a major coronary artery by the development of a thrombus or by an intimal hemorrhage with swelling of the arterial wall or by an embolus is termed coronary thrombosis

The injury or necrosis of a portion of heart muscle because of an interruption or curtailment of its blood supply is infarction of the myocardium or cardiac infarction

Acute coronary occlusion and myocardial infarction are not always associated Coronary occlusion may occur without an ensuing infarction and myocardial infarction may develop in the absence of an acute coronary occlusion The same symptoms are associated with myocardial infarction whether or not it is due to an acute coronary occlusion

There has been a progressive increase in the reported incidence of coronary occlusion This rising curve of reported mortality has not yet leveled off and it will not level off until clinical judgment in diagnosis supersedes diagnosis made by the electrocardiographic machine A person can die from some other cause despite the fact that he has a bad heart

There is great uncertainty however as to whether this reported increase is real or is due to a statistical mirage produced by increased knowledge and more accurate diagnosis of coronary thrombosis The increase in the span of life due to the reduction in diseases of childhood and other infections and to other advances in medical knowledge has brought a larger proportion of the population to the age

groups in which death from arteriosclerotic coronary thrombosis occurs There has always been a large number of individuals in the age group in which death from arteriosclerosis and coronary thrombosis is frequent and probably there has been but little change in the mortality rates from arteriosclerosis or its complications in the total numbers in this age group

Arteriosclerosis of the coronary arteries is usually responsible for acute coronary occlusion The occlusion is caused by intimal hemorrhage and subsequent formation of thrombus originating secondarily in an atherosclerotic plaque or in rupture of an arteriosclerotic abscess

There is a basic relationship of hypertension and arteriosclerosis to acute coronary occlusion This relationship seems to be founded on observation that hypertension intensifies or accelerates arteriosclerosis In some instances it seems that hypertension predisposes to the arteriosclerosis and as a result of arteriosclerosis there are an intimal hemorrhage in arteriosclerotic plaques formation of thrombus and coronary occlusion There are other conditions which seem to predispose to arteriosclerosis such as diabetes familial hypercholesterolemia with xanthomatosis myxedema and polycythemia to mention but a few Then there are those conditions which may seem to be important in the production of coronary occlusion and these may be separated into remote and immediate causes The *remote causes* may be enumerated (1) At least 9 of every 10 who have acute coronary thrombosis and myocardial infarction are between the ages of 40 years and 70 years When younger persons have acute coronary thrombosis death often occurs suddenly (2) Most of those who have coronary thrombosis are men (3) Occupation is secondarily an important predisposing factor for the reason that those who are temperamentally fitted for occupations requiring nervous tension are those who would have coronary thrombosis anyway (4) As statistical data accumulate racial predilections become less obvious (5) Morbid heredity appears to be a remote etiologic factor predisposing to the occurrence of general arteriosclerosis and therefore of coronary sclerosis and occlusion (6) Patients who have acute coronary thrombosis often are of short stocky stature have some obesity a thick short neck and a barrel shaped thorax These somatic attributes are inherited The disease occurs however in all the somatic types

Like the remote causes for coronary occlusion which have been enumerated some of the so called immediate causes have only slight demonstrable relationship to coronary occlusion The actual occlusion of a coronary artery precedes the symptoms of the attack often by days Efforts by the physician to discover the immediate causes by recording the events or activities immediately preceding an acute coronary occlusion are futile The history will reveal (1) Physical exertion may be indirectly related to the development of a myocardial infarct after an acute coronary occlusion has already occurred (2) Emotional strains must be interpreted in terms of their effect on the individual and his reaction to them Like physical exertion emotional stress cannot have a definite value assigned to it (3) Surgical operation may be followed by an acute myocardial infarction Those affected usually are more than 50 years old and often past 60 years Almost always there is evidence in these hearts that there was pre existing severe coronary arteriosclerosis and often a previous coronary occlusion (4) A patient who has had a certain attack of coronary occlusion confronts increased risk on subjection to surgical operation (5) There is no good evidence that the use even the excessive use of tobacco or overindulgence in alcoholic beverages contributes either remotely or immediately to the development of coronary artery diseases or acute coronary occlusion

PATHOLOGY A common cause of coronary thrombosis is the formation of a thrombus on an arteriosclerotic intimal plaque

Coronary occlusion occurs with equal frequency in the left anterior descending artery and the right circumflex artery Occlusion of the left circumflex coronary artery

occurs less frequently Cardiac enlargement or hypertrophy occurs in at least two thirds of those who have had cardiac infarction However the heart often remains of normal size after acute coronary occlusion Cardiac hypertrophy is seen pathologically and cardiac enlargement is observed clinically in many cases of coronary occlusion

SYMPTOMS In most of those who have not had angina pectoris premonitory thoracic pain precedes coronary occlusion by one to several days or weeks Those who have had angina pectoris realize from the beginning that the pain of coronary occlusion is different from the pain of angina pectoris It is distinctive because for the first time the pain commenced during sleep or rest and without apparent cause and it failed to subside promptly with rest or nitroglycerin After an hour or two of severity the pain of coronary occlusion may lessen perceptibly or subside entirely only to recur for hours or days at irregular intervals thereafter

In some patients there is intense pain for one or two hours after which the patient may feel and appear perfectly well Only electrocardiographic studies disclose that the pain was due to an acute myocardial infarction The pain of acute myocardial infarction like that of angina pectoris may extend to the shoulders and both upper extremities to the neck and jaw and to the interscapular region

After myocardial infarction the patient who has not previously had pain may become subject to paroxysms of angina pectoris These may occur as a result of slight activity meals or emotional states Other patients previously affected by attacks of angina pectoris may be completely relieved of their pain after they recover from the acute myocardial infarction

When myocardial infarction occurs without pain it is often characterized by a sudden development of weakness shock pulmonary edema or congestive heart failure However milder manifestation of myocardial infarction occurs during which the patient complains of transient cold sweats and weakness and of feeling sick dizzy faint or nauseated Vomiting and thirst are common

When shock occurs it comes soon after the onset of the pain and lasts from a few hours to one or two days or until death If the patient recovers from the shock state there is an awareness of pressure pain over the sternum or precordium pain or weakness in the arms or forearms or numbness in the wrist or fingers The persistence of shock is followed by congestive heart failure

In some patients who have myocardial infarction the first manifestation is an abrupt attack of pulmonary edema which appears suddenly and without warning Sometimes however it follows hours or days after premonitory substernal pain or recovery from the earlier stage of shock

A myocardial infarction which commences with massive pulmonary edema may terminate rapidly or if improvement supervenes there may remain left ventricular failure characterized by recurrent cardiac asthma orthopnea or dyspnea on slight exertion As time passes and subsequent infarctions occur evidences of right sided congestive heart failure ensue

EXAMINATION On examination there may not be present any abnormalities in appearance or behavior except that the patient complains of persistent thoracic pain Often there are evident weakness perspiration nausea or abdominal distention and breathlessness

In other patients who have myocardial infarction the physician may witness all the known emotional and physical acrobatics which have been and are practiced by mankind when recipients of torture and pain The emotional response of terror dominates Generally however the patient is listless or prostrated The responses to questions are brief There is evident desire on the part of the patient to be left alone and not be burdened with answers to questions or examinations The skin of the face is of ashen pallor and the lips and finger tips are faintly cyanotic A bluish red mottling of the skin (cutis marmorata) is common The hands and feet and the tip of the nose are cold and moist

In acute pulmonary edema the patient is propped up in bed gasping for breath and neck veins are engorged the lips and skin distinctly cyanotic There is a cough and the sputum is frothy and pink with blood If death is impending there is a copious foamy pink stained fluid bubbling from the mouth and sometimes from the nose Pulmonary edema often clears rapidly after an opiate or bloodletting (500 ml) It may persist intermittently for several days despite any known therapeutic measures

On percussion of the cardiac borders and palpation of the apical impulse in cardiac infarction there may be no evidence of cardiac enlargement In the presence of congestive heart failure the heart may be so enlarged that this is readily determined by percussion and palpation

Auscultation may reveal enfeebled heart sounds The first sound is usually faint and low pitched A soft blowing murmur at the apex is heard in instances of second or multiple infarction in which there is some degree of left ventricular dilatation and subsequent mitral insufficiency An aortic systolic murmur may be heard which is attributed to arteriosclerotic changes in the ascending aorta When hypertension persists despite the myocardial infarction the second aortic sound is accentuated or reduplicated In left ventricular failure accompanied by pulmonary hypertension the second pulmonic sound is accentuated and may be louder than the second aortic sound

Gallop rhythm and pericardial rub if present are best heard at or slightly medial to the apex or in the fourth interspace near the left sternal border The gallop rhythm is usually present when there is heart failure It most frequently appears in the first day or two after the infarction and disappears as cardiac function is improved and the pulse decreases

An intermittent pericardial rub is present in about one fifth of those who have cardiac infarction It may first be audible a few hours after the attack but most commonly on the second or third day In large infarcts or progessive myocardial infarction a pericardial rub persists continuously for many days It is usually heard along the left border of the sternum but it may be audible over a large part of the precordium The rub indicates an acute fibrinous pericarditis and not necessarily anterior infarction

The pulse rate often is between 100 and 110 per minute A sinus bradycardia with a pulse rate of 60 per minute may be present

In cardiac infarction there is first a rise in blood pressure This is followed by a fall in blood pressure which occurs sometimes after several hours but more often on the second day after the attack The decrease in blood pressure is by progressive decline and rarely it is delayed for a week In those in whom it results early the blood pressures may be less than 90 or 80 mm of mercury In about one third of the patients who have had hypertension before myocardial infarction the blood pressure remains within a normal range for at least many months thereafter In these there is no evidence of a diminished cardiac output Some never regain their hypertension

Acute cardiac infarction is accompanied by fever which begins within a few hours up to two days The temperature ranges from 100 to 102 F (37 7 to 38 8 C) or higher It usually reaches its peak on the third or fourth day and continues so for seven or eight days

An increase in the *leukocyte count* occurs on the second or third day only occasionally within hours after the infarction The count varies from 12 000 to 15 000 or 20 000 per cubic millimeter of blood The leukocytosis disappears by the end of a week There is an associated polynucleosis with a slight shift to the left A persistence of a high leukocyte count suggests the development of a complication such as pulmonary or other embolism bronchial pneumonia or progressive myocardial infarction

The rate of sedimentation of the erythrocytes is almost always increased after an acute myocardial infarction This test is of questionable diagnostic value since the sedimentation rate may be increased in almost every illness which affects the human race and at times when no illness can be assigned

Chemical examination of the blood may reveal hyperglycemia and azotemia which may prove to be transient unless present prior to the attack

The examination of the urine usually reveals no significant abnormality indicative of cardiac infarction Hematuria may appear as a consequence of renal infarction secondary to emboli arising from left ventricular mural thrombi

Electrocardiographic Changes in Acute Cardiac Infarction The electrocardiographic changes following cardiac infarction when studied early and by serial examination at brief intervals may be specifically diagnostic of the disease When coronary occlusion occurs without infarction there may be no significant electrocardiographic abnormality despite the occlusion

Electrocardiograms are often normal with slight coronary heart disease but in advanced disease they usually show changes in the Q and T waves and often in the S T segments They may show the presence of intraventricular block and low voltage uncommonly there is auriculoventricular block Abnormal left axis deviation is not common unless there is an associated hypertension

In the presence of coronary artery occlusion Q waves tend to appear or are exaggerated and the T waves may be deformed becoming low inverted or of unusual shape A few hours after onset a high origin of the S T segment from the descending limb of the R wave in lead I or lead III with a corresponding depression of this S T segment in the opposite lead that is lead III or lead I appears With this change in the S-T segments and T waves there appears a distinct Q wave in the QRS complex or an exaggeration of it if already present in lead I when the S T segment is elevated in lead I and in lead III when the S T segment is elevated in that lead In the apex lead (lead IV) the R wave tends to disappear the S T segment is much elevated in the early stages and the T wave becomes inverted in the Q_1T_1 type of electrocardiogram while the QRS and T waves remain unchanged or are exaggerated and the S T segment occasionally is definitely depressed at first in the Q_3T_3 type After a few days the S T segment begins to flatten out but there remains either in lead I or in lead III an inverted T wave The flattening or inversion of the T wave may persist or the deflection may gradually become normal again after weeks or months associated with recovery and repair of the infarct The abnormality of the Q waves often is permanent and remains as mute evidence of an old infarction

Infarction of the anterior wall of the left ventricle is generally attended by high origin and later inversion of T_1 development of Q_1 absence of R_4 and inverted T_4 while infarction of the posterior wall of the left ventricle is likely to be followed by high origin and later inversion of T_3 with development of a prominent Q_3 and a relatively normal lead IV The QRS and T waves in lead II may not be found altered but it is common for T to be flat or inverted during the course of the acute myocardial infarction and sometimes for months or years thereafter in either the anterior wall type of infarction of the posterior wall type more commonly in the anterior wall type the more marked and persistent the changes in lead I or lead III the more abnormal is T

Electrocardiography of a suspected coronary heart disease is not complete without at least one chest lead and several authorities believe that it is essential to take even as many as 6 and more recently 12 chest lead records especially in doubtful instances where there is need to explore to establish the lead point nearest to the area of damaged muscle Thus the routine cardiac apex lead (lead IV) may demonstrate or confirm by absence of the R waves or by elevation of the S T segments or by inversion of the T waves the presence of an infarct of the anterior wall of the left ventricle without bothering with any other precordial lead but a lateral wall or septal infarct may require leads further to the left or to the right and a posterior wall infarct may be shown only so it is said by an esophageal lead

In some instances the electrocardiographic changes may not be significant of acute myocardial infarction and thus do not disclose its situation This is particularly

true when there have been multiple infarcts Also the presence of a bundle branch block extensive pericarditis the administration of digitalis or a decidedly transverse or vertical heart each or in combination will obscure the electrocardiogram so that localization of the infarct is impossible

DIAGNOSIS OF ACUTE CORONARY OCCLUSION AND MYOCARDIAL INFARCTION In a middle aged or older man severe oppressive pain over the precordium or over the lower part of the sternum or more extensively over the anterior part of the thorax lasting at least a half hour and not relieved by the administration of nitrites indicates in most instances acute myocardial infarction

Myocardial infarction is suspected in a middle aged person when unexpectedly and without apparent cause there are severe prostration and collapse

When an aging person begins to experience attacks of nocturnal dyspnea or dyspnea on mild exertion and when these symptoms suddenly become more severe and there is perhaps enlargement of the liver engorgement of the peripheral veins or peripheral edema the possibility of a recent cardiac infarction is likely

In many instances of acute cardiac infarction the thoracic pain may have been mild and transient and will have been forgotten But if there should be the history of a sudden and unexplained extreme fatigability weakness attacks of dizziness or syncope indigestion short episodes of dyspnea and pallor or cyanosis palpitation and a cardiac arrhythmia myocardial infarction is a likely diagnostic consideration

There may be however certain electrocardiographic findings which in themselves justify the diagnosis The electrocardiographic studies are invaluable in the diagnosis of cardiac infarctions without pain

PROGNOSIS IN ACUTE CORONARY OCCLUSION AND MYOCARDIAL INFARCTION The duration of life when acute cardiac infarction occurs is unpredictable because from the instant the infarction occurs there is the constant threat of sudden unexpected death then and during convalescence and even thereafter during the lifetime of those who survive for years The mortality rate is 40 to 50 per cent within 4 to 6 weeks after onset

It is probable that if the milder instances of the disease were identified the mortality rate from first attacks of acute myocardial infarction would not exceed 1 in every 10 It seems that survival is the rule after the first occlusion or even after successive occlusions which have given no symptoms

Myocardial Infarction and Sudden Death Visceral embolism and peripheral embolism associated with myocardial infarctions are common complications and often the immediate cause of death These emboli in the following order of frequency affect the lungs the brain the kidneys the spleen the intestinal tract and the arteries The embolus often lodges at the bifurcation of a peripheral artery The commonest site is the femoral artery at the origin of the profunda femoris and the next commonest sites are the common iliac axillary and brachial and popliteal arteries and the aorta Peripheral embolism which does not cause immediate death is often characterized by acute and excruciating pain blanching of the affected area of skin loss of sensation coldness of the extremity loss of arterial pulsation and later loss of motion of the affected muscles However it is important to know that peripheral embolism may not be accompanied by pain In the course of serious or critical illnesses and particularly when the patient has severe pain embolism may occur without complaint and the patient be found dead by a member of the family or an attendant of the sick Among the causes of sudden death are massive pulmonary and cerebral embolisms

The weakened ventricular wall the result of infarction may permit the formation of ventricular aneurysm

Sudden death may be caused by the rupture of a cardiac aneurysm Sudden death from rupture of the heart in the absence of cardiac aneurysm is of relatively common occurrence in the first few weeks after acute myocardial infarction

Ventricular fibrillation is a common cause of death after infarction

Silent Coronary Occlusion Painless coronary occlusion may occur under circumstances which are unpredictable in patients who have not had prior known heart or circulatory disease The cause however is usually an atherosclerosis of the coronary arteries Although the attack does not cause pain it seems that these patients have difficulty in swallowing and are subject to choking gagging and some of them dyspnea These manifestations may be considered as equivalents of pain

Hypertensive Arteriosclerotic Heart Disease Hypertension arteriosclerosis atherosclerosis and hypertensive heart disease may be related to bodily build and constitution These may be more frequently present in the obese and occur with some predilection for the short short necked stocky individual But are not short stocky individuals predominant in the general population?

Hypertension arteriosclerosis and atherosclerosis are uncommon before the age of 20 years Before the age of 20 years hypertension occurs chiefly with co arctation of the aorta with acute or chronic glomerulonephritis and rarely with endocrine tumors Thereafter it is encountered in 20 to 25 per cent of the general population and in 40 to 60 per cent of those more than the age of 50 years

Atherosclerosis occurring before the age of 20 years is usually associated with diabetes mellitus or xanthomatosis of the congenital variety or a combination of diabetes and xanthomatosis Atherosclerosis in the youth may or may not be asso ciated with hypertension and heart disease

Compensated Hypertensive Heart Disease In hypertensive heart disease the heart is hypertrophied and capable of doing its work (compensated) or it is unable to do its full work (decompensated) Unless increased or excessive demands are made on the heart in compensated hypertensive heart disease symptoms may long be delayed When symptoms do make their appearance they are those of left ventricular heart failure

In the compensated stage of hypertensive heart disease the left ventricle is enlarged Left ventricular hypertrophy is suggested by a forceful apical impulse due partly to rotation of the apex nearer the thoracic wall and partly to a more forceful systolic contraction The second aortic sound is accentuated Compensated hypertensive heart disease does not in itself cause murmurs but an apical or basal systolic murmur may be caused by mitral or aortic sclerosis and calcification In elderly patients who have severe hypertension and pronounced dilatation of the ascending aorta so called arteriosclerotic aortic diastolic murmur is heard occa sionally

Elongation of the aorta as it loses its elasticity attended by palpable pulsations in the episternal notch with concomitant elevation of the subclavian carotid and innominate arteries gives rise to strong arterial pulsations in the neck The carotid arteries are occasionally distorted and kinked in such a manner as to produce localized pulsating bulges which may resemble an aneurysm A congenitally long carotid artery may cause a comparable pulsation

Increased pulsation of the aorta is common in hypertension In cases of long standing hypertension and aortic arteriosclerosis elongation and elevation of the aortic shadow are frequently observed roentgenoscopically

In one fourth to one half of those who have moderate hypertension or arterio sclerosis or a combination of these conditions the electrocardiogram reveals no abnormality Often there is only a left axis deviation but this may be absent even when the severe hypertension seen in the malignant essential hypertension is present

Decompensated Hypertensive Heart Disease Cardiac decompensation in the hypertensive heart is almost always associated with advancing coronary arteriosclerosis Heart failure involves the left ventricle but later there is evidence of congestion in the systemic circulation as well as in the pulmonary circulation

The hypertensive individual is characterized as a hyperreactor with exaggerated response to various influences such as cold and epinephrine

PATHOLOGY In hypertensive heart disease there is hypertrophy of the left ventricle When heart failure comes to those who have hypertension there are present both dilatation and hypertrophy The left ventricle is primarily affected The right chambers too may be dilated and hypertrophied as right heart failure is inevitable and when it develops it persists

Coronary arteriosclerosis with or without severe narrowing and with or without occlusion and infarction may be present and may be the immediate cause of heart failure in hypertension Ischemic necrosis occurs in the hearts of thos who have malignant hypertension

It is during the stage of acute heart failure that pronounced cardiac enlargement occurs A systolic murmur if present is often due to functional mitral insufficiency Gallop (3 1) rhythms and pulsus alternans are common With pulmonary congestion the second pulmonic sound may be louder than the second aortic sound despite systemic arterial hypertension Auricular fibrillation occurs in 1 of every 4 of those who have hypertensive heart disease Frequently it seems to precipitate heart failure

The lungs contain water and there is often edema of the dependent parts

The combination of diastolic hypertension and left ventricular hypertrophy together with evidence of angina pectoris or coronary occlusion and myocardial infarction and congestive heart failure or electrocardiographic signs of intraventricular conduction disturbances or both should permit of a diagnosis of hypertensive and arteriosclerotic heart disease if aortic stenosis is excluded

Survival for 10 to 20 years and even 30 years is not unusual In the malignant phase of hypertension characterized by diastolic pressure of 130 mm of mercury or more neuroretinitis and progressive renal insufficiency the prognosis is grave

THE ENDOCARDIUM

Endocarditis The term endocarditis is used with the appropriate adjective (bacterial viral spirochetal rickettsial or mycotic) in connection with diseases of the endocardium due to a known causative organism The term nonbacterial endocarditis designates an endocarditis usually associated with rheumatic fever or some general disease perhaps the result of a bacterial sensitization reaction such as rheumatic endocarditis and the endocarditis of Libman Sacks Nonbacterial thrombotic endocarditis occurs in terminal or cachectic conditions Degenerative alterations of the endocardium are common These include endocardial sclerosis calcification ossification fatty degeneration and amyloidosis

The presence of superficial excrescences verrucae on the surface of the valvular or occasionally on the parietal endocardium is the obvious feature of an endocarditis Valvular endocarditis may occur without the formation of verrucae or verrucae may be present on one or more of the valves while other valves are free

Acute Bacterial Endocarditis Bacterial endocarditis occurs in acute and subacute forms The acute forms of disease are mentioned in association with descriptions of the causative organisms The disease occurs when there is bacterial invasion of the blood stream with localization of the bacteria on the endocardium Once the bacteria are seated on the endocardium there are persistent bacteremia toxemia and embolization due to the bacterial vegetations breaking away from the endocardium Acute bacterial endocarditis is characterized by having been preceded by a local or a general infection Many of these patients have had previous endocardial disease Many of them too have normal hearts at the beginning of the acute endocarditis

Acute bacterial endocarditis is commonly due to hemolytic streptococci pneumococci or *Staphylococcus aureus* The hemolytic streptococci belong serologically to Lancefield s group A but occasional instances due to streptococci of group C or B subsequent

to abortion or delivery occur Pneumococci responsible for acute bacterial endocarditis are usually aerobic but may be anaerobic

Among a variety of other organisms responsible for acute bacterial endocarditis are *Neisseria gonorrhoeae* *Neisseria meningitidis* *Escherichia coli* *Pseudomonas aeruginosa* (*Bacillus pyocyaneus*) *Klebsiella pneumoniae* (Friedlander s bacillus) *Actinomyces bovis* *Micrococcus rugatus* spirilla and *Bacillus anthracis* Many of this latter group may produce either an acute or a subacute bacterial endocarditis

Age sex and race do not appear to be significant predisposing factors

In acute bacterial endocarditis the vegetations may be superimposed on thickened scarred vascularized rheumatic valves or on congenital cardiac lesions or they may be on valves with little minimal or no previous deformity The pneumococcal and gonococcal infections often occur on previously normal valves and they affect the aortic valve more frequently than do the other organisms Vegetations on the right side of the heart are caused most often by the gonococcus

Ulceration is a feature of the acute form of the disease valvular destruction being especially severe in the pneumococcal and gonococcal infections The interventricular septum and rarely the cardiac wall may be perforated An accompanying pericarditis is a common finding

The *spleen* is enlarged and may contain small abscesses The *liver* may disclose focal or diffuse areas of necrosis and degeneration The *kidneys* often present hemorrhages infarcts and sometimes small or large abscesses One large abscess renal carbuncle may occur in staphylococcal septicemia Multiple renal abscesses are characteristic of this septicemia There may be a focal or acute diffuse glomerulonephritis The *eye* may be affected by panophthalmitis but more commonly there is simply a choroiditis

The general symptoms are those of the primary infection which has originated the bacteremia During the course of such an infection a chill and fever may signify the beginning of the septicemia The fever then persists despite the disappearance of the original focus of infection which seems to have healed In other instances during the course of an infection there are chills profuse sweats marked weakness and various cerebral and psychic disturbances embolic and vascular symptoms consisting of vascular occlusions in parenchymatous organs or in the extremities and cutaneous lesions The vascular occlusions may affect the brain producing hemiplegia There may be occlusion of peripheral arteries Pulmonary infarctions causing hemoptysis are common All of these complications give their respective localizing symptoms

On examination cutaneous manifestations consisting of petechiae hemorrhages and erythematous bullous and purulent lesions may be evident The spleen may be palpable The other physical findings are those of the original focus of infection This focus may not be evident

The heart rarely contains any physical abnormalities which are of significance There may be a systolic murmur or an increase in intensity of one which may have been present There are a rapidly developing anemia a leukocytosis and perhaps a positive blood culture

In acute bacterial endocarditis the culture of organisms from the blood stream confirms only the diagnosis of a bacteremia The appearance of successive generations of white centered cutaneous petechiae in a patient who has a proved bacteremia is very suggestive that the endocardium is likewise affected If on ophthalmoscopic examination petechiae are present in the choroid or if a panophthalmos develops acute bacterial endocarditis is a likely diagnosis

Subacute Bacterial Endocarditis In 9 of every 10 who have subacute bacterial endocarditis the causative organism is a nonhemolytic streptococcus The causative organisms in the rest of those who have subacute bacterial endocarditis comprise a great variety of microorganisms Almost any organism capable of producing septicemia may cause subacute bacterial endocarditis

Subacute bacterial endocarditis almost always affects a heart which has valvular defects usually acquired from an earlier rheumatic fever A few patients in the

younger age groups have congenital cardiovascular anomalies which predispose to subacute bacterial endocarditis

Subacute bacterial endocarditis often develops after some inconsequential event such as the extraction of a tooth or the removal of tonsils or a bad cold especially in those who have rheumatic or congenital cardiac valvular defects

The highest incidence of this disease is from 20 through 29 years of age but subacute bacterial endocarditis may occur at any age from early childhood to late in life There is a somewhat higher incidence among men than among women

PATHOLOGY The vegetations are usually situated on the leaflets of the mitral valve but may affect any or all of the valves

The myocardial lesions consist of small infarctions bacterial embolization of arterioles and capillaries necrosis of the walls of these small vessels and surrounding inflammatory reactions

In subacute bacterial endocarditis often the *kidneys* may present diffuse petechial hemorrhages which produce depressed yellowish sharply demarcated lesions of the cortex indicating the sites of infarction The *spleen* is almost invariably enlarged and contains infarcts

Lesions of the *brain* are frequent and widespread There is often a diffuse meningo encephalitis due to disseminated bacterial embolization of arterioles and capillaries Embolic lesions of the large cerebral arteries are common and in some cases prove fatal Bacterial emboli lead to mycotic aneurysms and thrombosis Cerebral intraventricular or subarachnoid hemorrhage may result from rupture of a mycotic aneurysm Encephalomalacia is a consequence of vascular (embolic) occlusion Occasionally small or large cerebral abscesses are formed

In the lungs congestion pulmonary emboli and infarctions and bronchopneumonia may be present The liver is enlarged and engorged owing to chronic passive congestion The stomach and the intestines almost always are involved during the course of subacute bacterial endocarditis the involvement originates from embolic phenomena Bacterial emboli also affect the large or small arteries of the extremities causing circulatory disturbances and in some gangrene

SYMPTOMS The symptoms of subacute bacterial endocarditis are referable to the general infection to the embolic and vascular phenomena and to the affected heart The onset is usually insidious It may commence acutely however with chills and high fever An acute onset is generally due to embolus in a cerebral an abdominal or a peripheral vessel

At the onset fever is the most constant of all symptoms It may be continuous remittent intermittent or septic The temperature is rarely more than 103 F (39 4 C) the usual range is between 100 and 102 F (37 7 and 38 8 C) Periods of apyrexia lasting for weeks at a time often supervene Shaking chills or chilly sensations are common During the chill the temperature is likely to reach 104 to 105 F (40 to 40 5 C) with quick rise and sudden fall of the temperature Profuse night sweats even in the absence of known fever are common Pallor weakness headaches tenderness over the long bones and the sternum breathlessness and easy fatigability are common subjects for complaint

Anorexia abdominal pain nausea and vomiting are common and diarrhea may be severe For weeks in the beginning of the disease diarrhea and fever may be the only symptoms

Splenic infarction may be characterized by sharp sudden pain in the left side of the hypochondrium or the lower part of the left axilla which may extend to the left shoulder or precordium There may be local soreness and hypersensitivity of the skin over the splenic region

Cerebral or spinal cord infarction may be manifest by the sudden occurrence of hemiplegia or monoplegia In the symptomatology of subacute bacterial endocarditis it is well to be aware that many of the emboli reach small arteries and cause localized symptoms more frequently than they reach the arteries to vital organs and produce alarming symptoms

EXAMINATION The *skin* is pale and often there are petechiae The petechiae are most frequently and best observed in the conjunctiva especially of the lower lid in the oral mucosa and on the soft and hard palates in the fundus in the skin about the clavicles and in the lower extremities Some petechiae may have white centers Petechiae occur in successive generations Each generation disappears after several days to be succeeded by another at some future date

Tender cutaneous lesions which are raised swollen patches from a few milli meters to 0 5 cm in diameter bluish or pink or red occur in the pads of the fingers or toes in the thenar or hypothenar eminences on the soles of the feet or between the toes These lesions are always tender It seems that these tender nodes when present are pathognomonic of subacute bacterial endocarditis

The cardiac findings are those of the existing valvular disease or congenital malformation In the beginning of subacute bacterial endocarditis valvular lesions may be present of which the patient is unaware As the heart is repeatedly examined during the progress of the disease there are changes in the character and sites of murmurs or new murmurs develop A new murmur rough in tone which appears suddenly may be due to rupture of a chorda tendinea or perforation of a valve cusp

Jaundice may be present and the liver may or may not be palpable The jaundice may be associated with pulmonary infarction or with gonorrheal endocarditis The spleen is palpable in at least two thirds of those who have subacute bacterial endo carditis

Clubbing of the fingers and of the toes is frequent

Sudden blindness and ophthalmitis may result from embolism of the central retinal artery Visual field defects and fever when of sudden onset may be suspected as being the result of subacute bacterial endocarditis Retinal hemorrhages may be present (see Diseases of the Eye Chapter 4)

Anemia is a common feature of the disease Leukocytosis is not characteristically present unless there is some complication The sedimentation rate of erythrocytes seems to be accelerated in subacute bacterial endocarditis Polymorphonuclear leukocytosis is slight if present at all Blood cultures are found to be positive in 9 of 10 who have the disease Cultures may have to be repeated before positive ones are obtained

The urine may contain blood and albumin especially after infarctions of the kidneys There may be a focal glomerulonephritis present

DIAGNOSIS The diagnosis of subacute bacterial endocarditis is based on the presence of (1) a positive blood culture (2) embolic phenomena (3) a febrile course and (4) a valvular defect or congenital cardiovascular lesion The diagnosis of subacute bacterial endocarditis should be assumed as most probable whenever a patient who has an organic cardiac murmur experiences fever without apparent cause for more than one week

Blood cultures have acquired additional importance above that of diagnosis because beneficial treatment is most likely when the causative organism is identified and its sensitivity to individual antibiotics is determined

VALVULAR HEART DISEASE

There are two types of valves in the heart the bicuspid (mitral) and the tri cuspid between the atria (auricles) and the ventricles and the two sets of semilunar valves at the entrance of the pulmonary artery and aorta

The *bicuspid* or *mitral valve* is the most important and the most deeply seated It is situated under the edge of the left border of the sternum opposite the fourth costal cartilage It separates the left atrium and ventricle and lies nearly transversely

The *tricuspid valve* is situated under the middle of the sternum opposite the fourth intercostal space It runs obliquely downward and to the right from the third left inter costal space to the fifth right costal cartilage It separates the right atrium and ventricle

The *pulmonary semilunar valve* lies opposite the sternal end of the third left costal cartilage It is the most superficial valve and the one highest up on the sternum It prevents regurgitation of the blood into the right ventricle from the lungs

The *aortic semilunar valve* is situated under the left side of the sternum about level with the lower edge of the third costal cartilage

Mitral Valvular Disease Rheumatic endocarditis of rheumatic fever is responsible for almost all mitral valvular disease Mitral valvular disease occurs as (1) mitral insufficiency and (2) mitral stenosis

Mitral Insufficiency This form of disease occurs as the result of imperfect closure of the valves The disease is called also organic mitral insufficiency and relative mitral insufficiency

In rheumatic fever there is first an organic mitral insufficiency Other causes for organic mitral insufficiency are bacterial endocarditis traumatic rupture of a cusp or of the chordae tendineae spontaneous rupture of the chordae and rupture of a papillary muscle

Organic mitral insufficiency is consequent to incomplete closure of the mitral orifice which results from imperfect coaptation of the valve cusps because of scarring thickening shortening and deformity In others there are fusion and shortening of the chordae which fix the cusps and restrain their apposition so as to prevent closure of the valvular orifice In still others there exist inflammation and scarring of the mitral ring which dilate the orifice and interfere with the muscular systolic shrinkage in the size of the ring

Rupture of the mitral valve may cause mitral insufficiency by perforation or tear of a cusp or by rupture of chordae or a papillary muscle Spontaneous rupture of chordae tendineae usually of previously diseased valves may occur

Relative or functional mitral insufficiency occurs commonly as a result of great dilatation of the left ventricle in association with left ventricular failure Imperfect valvular closure under such circumstances is due to dilatation of the mitral ring and to retraction of the cusps by the chordae and papillary muscles Relative or organic mitral insufficiency may be caused by any of the etiologic causes of heart failure these are mainly rheumatic fever and arteriosclerosis

The left atrium dilates and hypertrophies in both mitral insufficiency and mitral stenosis It may attain a volume of 2 or 3 liters In contrast an advanced mitral stenosis may be present without atrial enlargement if the heart is generally enlarged

In hearts with predominant mitral stenosis the left ventricle is of normal size or may be smaller than normal or atrophic

Symptoms Many patients who have mitral insufficiency enjoy a normal life Compensation of mitral insufficiency is effected by dilatation and hypertrophy of both the left atrium and the left ventricle When symptoms appear they are those of congestive heart failure

Examination On physical examination there may be only a loud harsh or blowing systolic murmur near the apex with varying degrees of transmission toward the axilla A murmur which is not transmitted to the axilla is usually indistinguishable from insignificant functional murmurs

If roentgenoscopic examination reveals an enlargement of the left atrium and the left ventricle the significance of the murmur is thus assured

If there is a rupture of the mitral chordae tendineae the systolic murmur is loud and is associated with a thrill There may also be an associated apical diastolic murmur

Heart failure resulting from mitral insufficiency is rare If heart failure should be present it results from an active rheumatic myocarditis or an unusual strain such as may attend gestation

The electrocardiogram in mitral insufficiency shows a tendency to left axis deviation The P waves may be tall and notched

DIAGNOSIS The diagnosis of mitral insufficiency is based on the presence of an apical systolic murmur and enlargement of the left atrium and ventricle and usually a history of rheumatic fever or chorea

The diagnosis of mitral insufficiency due to nonrheumatic causes is based on the sudden development of a loud apical systolic murmur and thrill in the course of bacterial endocarditis or myocardial infarction after thoracic injury or after severe exertion

The presence of an apical systolic murmur in a patient who has hypertension or coronary artery disease and heart failure which disappears with improvement of the circulatory status confirms the functional nature of the disturbance

Calcification of the mitral cusps almost always occurs in organic rheumatic mitral insufficiency or stenosis

If there is no more evidence of heart disease than a murmur there is no certain method of differentiating whether the murmur is organic or functional in origin Functional murmurs are more commonly present than organic murmurs and are the loudest in the pulmonic region transmit poorly and are faint in intensity Cardio respiratory murmurs disappear when the breath is held

Mitral Stenosis Mitral stenosis is due mainly to rheumatic endocarditis but occasionally it is a congenital defect due either to anomalous development or to fetal endocarditis Rheumatic mitral valvular disease occurs much more frequently among girls and women than among boys and men

In mitral stenosis the mitral orifice is obstructed and too little blood flows from the left atrium into the left ventricle during ventricular diastole and consequently into the aorta in ventricular systole Hypertrophy and dilatation of the left atrium develop and increased pressure in the pulmonary circulation and hypertrophy of the right ven tricle ensue The right ventricle dilates when it becomes inadequate for the work required of it

Thrombi arising in the auricles occur at any stage of mitral stenosis, but they are especially frequent after right heart failure and development of atrial fibrillation In the absence of right heart failure and consequent dilatation of the right atrium the thrombi are usually confined to the left atrium

Embolization of the kidneys spleen hepatic artery mesenteric artery and peripheral arteries occurs Embolization of the aorta with formation of a thrombus at its bifurcation may cause sudden sharp pain in the abdomen lower part of the back and the extremi ties loss of pulsation and warmth of the lower extremities and shock More localized symptoms occur with embolization of the popliteal femoral or brachial arteries Gan grene may result Subsequent emboli in other parts of the body may eventually cause death Extensive infarction of the lungs and spleen may occur in which minimal or no recognizable symptoms are present

A sudden hemiplegia may occur in the presence of a well compensated mitral stenosis Sometimes these hemiplegias are relatively benign in that they clear up with but slight disability only to be repeated Occlusion of a small cerebral artery may occur followed by only a focal paralysis or isolated aphasia

SYMPTOMS Often patients who have mitral stenosis have no cardiac symptoms and do not have a known history of rheumatic fever or chorea There is no mani fest evidence of a decrease in the cardiac reserve A normal life is and has been pursued Some of these men and women will be aged but there is only a minimal degree of arteriosclerosis such a slight degree that there is not the least suspicion that the cardiac signs are originating from this source

Some of these patients however are not so fortunate for there comes a time when cardiac compensation is imperfect The lungs become congested and attempts at unusual degrees of bodily exertion result in dyspnea

Mild degrees of disturbance of gaseous exchange in the lungs for instance at the stage of left atrial failure may cause dyspnea and cyanosis A mild degree of cyanosis produces the so called mitral facies which is characterized by a cyanotic flush of the skin over the malar prominences and a dusky cyanosis of the lips which is most

evident after exertion Severe degrees of cyanosis are present in long standing instances of mitral stenosis

Dyspnea on exertion is the commonest complaint of the patient who has mitral stenosis Many patients who have mitral stenosis become so accustomed to a mild degree of chronic dyspnea that they are scarcely aware of its presence

Palpitation and precordial distress are early and constant symptoms The palpitation is especially disturbing at night the patient often attributes it to indigestion The onset of auricular fibrillation may be described by the patient as palpitation Fibrillation however is usually established and associated with a slow ventricular rate for a long time without the patient s being aware of its presence

A cough worse at night and after physical exertion than at other times may be an annoying symptom Secondary bronchial and pulmonary infections are prone to complicate pulmonary congestion in mitral stenosis and these may be partially responsible for the cough

Hemoptysis may be the initial and only complaint It may occur suddenly and be of considerable severity Hemoptysis has been attributed to the rupture of small congested vessels in the lungs to the rupture of varices of the bronchial veins and to pulmonary embolization and infarction The ultimate prognosis in those who have hemoptysis is poor

Acute pulmonary edema is much less frequent in mitral stenosis than in hypertensive heart disease or in aortic valvular lesions Pulmonary edema follows sudden or severe exertion surgical operations pregnancy childbirth attacks of paroxysmal tachycardia or intravenous administration of physiologic saline solution

Angina pectoris when present is due to an associated aortic or coronary artery disease rather than to the mitral stenosis It is rarely present

Paralysis of the left recurrent (laryngeal) nerve may occur as the result of an enlarged left atrium pressing on the nerve The hoarseness may be intermittent at first but may progress to complete aphonia

Examination In compensated mitral stenosis the pulse is generally normal Fibrillation extrasystoles and dropped beats are frequent

Auricular fibrillation is present in about half of those who have mitral stenosis The fibrillation may be paroxysmal at first but becomes persistent It may be an accompaniment of right heart failure or may actually precipitate it However auricular fibrillation with slow ventricular rate is often compatible with years of comfortable life

The blood pressure in mitral stenosis is usually within normal limits However a hypertension may be present in some instances

In children the precordium may bulge Often there are visible pulsations aside from those of the precordium in the epigastrium and to the right of the sternum

A late diastolic (presystolic) thrill may be palpated as a vibrating sensation on the surface of the precordium in the region of the apical impulse A palpable shock may accompany the first sound at the apex and the second sound in the pulmonic region The apical impulse is usually felt at its normal site After experience has been gained a presumptive diagnosis of mitral stenosis can sometimes be made from palpation alone

The total area of cardiac dullness to percussion is often increased The increase in the area of cardiac dullness is along the left sternal border at the level of the third interspace and fourth rib This increase in dullness at the base of the heart is present in both mitral stenosis and insufficiency

The first sound at the apex is short loud and booming A similar booming first sound may be heard in hyperthyroidism

The second sound heard over the pulmonic area is accentuated and may be accompanied by a palpable shock The pulmonic second sound may be duplicated as well as accentuated

There is a localized apical diastolic murmur This may be late in diastole (presystolic) or it may fill most or all of diastole It may occur in the early or middle period of diastole The diastolic murmur presumably arises from the noise made by rapid blood flow past the mitral obstruction into the left ventricle The time in diastole at which this murmur occurs is that at which the pressure difference between the left atrium and the left ventricle is excessive This pressure difference is accentuated when the left atrium undergoes compensatory dilation and hyper trophy

The late diastolic (presystolic) murmur in mitral stenosis is of a low tone with a crescendo accentuation merging into an abnormally loud first sound The murmur is well circumscribed at or near the apex beat This diastolic (presystolic) murmur may be accentuated by a left lateral position after exercise has quickened the heart rate This murmur is absent until the obstruction of mitral stenosis is significant and it disappears at the onset of auricular fibrillation and heart failure

A diastolic murmur (presystolic in time) may be heard in aortic insufficiency severe anemia acute nephritis with hypertension and left heart failure as well as in mitral stenosis

An apical systolic murmur is frequently present owing to significant mitral regurgitation

On occasion a soft blowing diastolic murmur known as the Graham Steell murmur is heard over the pulmonic region It is probably the result of a functional insufficiency of the pulmonic valve due to pulmonary artery hypertension and con sequent dilatation of that vessel and of the pulmonic orifice The Graham Steell mur mur is distinguished from the diastolic murmur of aortic insufficiency by differences in the roentgenologic appearance of the heart and by distinctive peripheral signs

The roentgenologic appearance of the heart in mitral stenosis varies with the stage or severity of the lesion

Calcification occurring on the mitral valves or on the annulus fibrosus in the aged is clinically unimportant Calcification of the mitral ring or cusps may be associated with aortic stenosis also the result of calcification Deposits of calcium may be present in the fibrous septum which in time affects the bundle of His and thus heart block or other disturbances of conduction ensue

In the *electrocardiogram* in mitral stenosis the P waves in leads I and II are often widened and notched and may be of increased voltage There is usually right axis deviation with evidence of right ventricular strain Occasionally there is evi dence of right bundle branch block Auricular fibrillation is frequent

Diagnosis The diagnosis of mitral stenosis is based on a history of rheumatic fever on the foregoing described physical signs and on roentgenologic evidence of a large left atrium The diagnosis is more certain if the characteristic diastolic (pre systolic) murmur is heard A sharp snappy first sound at the apex following the second sound at the apex and a diastolic thrill further support the diagnostic evi dence Electrocardiographic records showing right axis deviation large notched P waves and auricular fibrillation also suggest mitral stenosis especially if there is a history of rheumatic fever

Aortic Valvular Disease ***Aortic Insufficiency*** Insufficiency of the aortic valve results from rheumatic fever syphilis bacterial endocarditis trauma severe hypertension and aortic arteriosclerosis and dissecting aneurysm of the aorta Aortic insufficiency in patients less than 35 years old is almost exclusively rheumatic in origin and is combined with mitral valvular disease

Some blood regurgitates from the aorta into the left ventricle during diastole Since in diastole the ventricle also receives its normal amount from the left atrium its cavity dilates At each systole a large amount of blood is delivered into the aorta and the wall of the left ventricle hypertrophies

The heart in aortic insufficiency is very large (oxheart or cor bovinum) weighing

from 500 gm to 1 000 gm The enlargement involves the left ventricle and interventricular septum

SYMPTOMS If the left ventricle is normal and the mitral valve sufficient a normal span of life may be expected without symptoms In those who have compensated aortic insufficiency the symptoms are often subjective and referable to the forceful ventricular contractions For instance when the patient lies on the left side the audible beat of the heart and cervical vessels is disturbing The conventional crossed legs may be uncomfortable from the pulsating popliteal artery Vascular pulsation of the uvula may cause an uncomfortable tickling in the throat

Those who have rheumatic aortic insufficiency may have nocturnal angina pectoris accompanied by a rise in blood pressure increased pulse and respiratory rate palpitations sweating and flushing of the face In aortic insufficiency of syphilitic origin angina pectoris is often due to the frequency associated stenosis of the coronary ostia or to conventional atheromatous coronary heart disease

A rapidly developing breathlessness on exertion is the first symptom of left heart failure in aortic insufficiency The dyspnea may be paroxysmal appearing suddenly while the patient is at rest as well as during activity Paroxysmal dyspnea of short duration is characteristic of syphilitic aortic disease Orthopnea is soon added

EXAMINATION The pulse rate is normal in compensated aortic insufficiency The pulse wave in aortic insufficiency rises and falls rapidly (Corrigan pulse) To find the so called water hammer pulse it is well that the patient s arm be elevated above the head and the pulse palpated in this position

In aortic insufficiency there are marked pulsations in the vessels of the neck in the temporal arteries and in other large vessels which may impart a to and fro motion to the entire head or to the leg when crossed over the opposite knee Ophthalmoscopic examination reveals pulsations of the retinal arteries as well as of the veins

The capillary pulse may be observed in a normal person when the skin is warm In sickness the phenomenon is observed in fever anemia hyperthyroidism and aortic insufficiency Capillary pulsation may be elicited in several ways (1) by observing the nail bed or skin at the root of the nail while pressure is made on the tip of the nail (2) by observing the edge of the blanched region when the mucous membrane of the lip is compressed by a glass slide (3) by observing the earlobe or finger tip while it is being transilluminated by a weak source of light

The apical impulse is forceful and is situated to the left of and below its normal site It may be seen or felt in the left axilla A diffuse systolic depression of the anterior thoracic wall medial to the forceful apical beat is caused by the wide excursions of the contracting ventricle within the closed thoracic cage

Percussion reveals an enlargement of the heart to the left and downward There is a soft high pitched diastolic murmur In syphilitic aortic insufficiency it is often louder and lower pitched than in the rheumatic form of the disease In traumatic aortic insufficiency the murmur is often extremely loud and has a pleasant quality It starts immediately after the second sound It is best heard in the second right intercostal space or along the left border of the sternum in the third or fourth interspace

A low diastolic blood pressure is regularly present in aortic insufficiency (60 mm of mercury or lower) The systolic pressure is normal or increased and thus the pulse pressure is increased

The diastolic murmur of aortic insufficiency is often a difficult heart murmur to hear In searching for the murmur ask the patient to stand to bend slightly forward or assume the crawling position and hold the breath intermittently during auscultation A systolic murmur situated at the base of the heart when present may be helpful for purposes of timing the murmurs

A late diastolic apical murmur (Austin Flint murmur) occasionally is heard in aortic insufficiency when there is left ventricular failure This murmur is identical with the late diastolic (presystolic) murmur of mitral stenosis The mitral valve is normal however in those who have an Austin Flint murmur

On auscultation over the femoral artery a booming sound synchronous with each pulse through the vessel may be heard This sound has been described as resembling a pistol shot In some instances a double sound (Traube s double tone) is heard in aortic insufficiency instead of the single pistol shot sound

A systolic murmur is present over the femoral artery in normal persons if the vessel is slightly compressed by the bell of the stethoscope In aortic insufficiency a diastolic as well as a systolic murmur is audible over the compressed vessel because there is a rapid diastolic regurgitation toward the heart as well as systolic forward passage toward the periphery This double murmur (Duroziez s sign) is not limited to aortic insufficiency but may also be present in hyperthyroidism febrile states and anemia

DIAGNOSIS A diagnosis of aortic insufficiency can be made when there are enlargement of the left ventricle a high pitched but often scarcely audible diastolic *murmur over the aortic region or along the left border of the sternum and when* present peripheral circulatory phenomena There is an increased pulse pressure obtained by use of the sphygmomanometer

Cardiac failure is an integral part of the course of aortic insufficiency and there may be no significant symptoms before failure occurs Sudden death is frequent in this form of valvular heart disease

Aortic Stenosis Rheumatic fever is the chief cause of aortic stenosis before the age of 50 years Aortic stenosis after the age of 50 years results from atherosclerosis

The heart in all forms of aortic stenosis is hypertrophied In the smaller hearts the hypertrophy is confined to the left ventricle In the larger hearts dilatation and hypertrophy involve the right ventricle as well as the left ventricle and often the atria

Complete heart block bundle branch block intraventricular conduction defects and delayed auriculoventricular conduction frequently are present and result from an extension of the calcific process from the aortic valve to the fibrous septum

SYMPTOMS Compensated aortic stenosis is asymptomatic When valvular obstruction develops then symptoms commence

Cardiac pain is present in 1 or 2 of every 10 who have aortic stenosis Angina pectoris is more frequently suffered by those who have aortic stenosis than by those who have other valvular heart lesions

Dizziness faintness and syncope associated with exertion or change of position may be the subject of complaint Convulsive disorders may occur as a part of the symptom complex of aortic insufficiency

Dyspnea on exertion is the first symptom of left heart failure in aortic stenosis It may be present for months or even a year or two before the other symptoms commence In time nocturnal paroxysmal dyspnea (cardiac asthma) supervenes Insomnia nightmares cough and expectoration occasional hemoptysis and orthopnea are symptoms of heart failure and are common in aortic stenosis

EXAMINATION The pulse in advanced aortic stenosis is of small amplitude rises slowly and falls away slowly (pulsus parvus et tardus) Its slow maintained elevation imparts to it the form of a plateau There is a delay in arrival of this slow small pulse at the wrist

The blood pressure in aortic stenosis may be within normal limits The systolic pressure may be slightly diminished and the diastolic slightly elevated with a resultant small pulse pressure even though these pressures are still within the range of the normal limits The systolic pressure on the average is increased

Palpation often reveals a systolic thrill at the base of the heart to the right of

the sternum or along the right cervical vessels This thrill is best felt if the patient sits or stands and bends forward

Over the aortic region there is a loud long systolic murmur which is rough in quality and is transmitted to the neck and to the left axilla The murmur may be heard downward along the sternum toward the apex and occasionally in the left infrascapular region in the back A diastolic murmur over the base of the heart often is present

The second sound over the aortic area is normal or is heard with difficulty or it is absent

The presence of effusions in the serous cavities particularly hydrothorax cyanosis weakness loss of weight and emaciation portend an impending disaster Sudden death is common in this disease

Roentgenologic examination reveals that in size the heart is either normal or diminished

The electrocardiogram may reveal normal conditions or may show only a left axis deviation if there is no associated mitral stenosis However there may be permanent or transient conduction disturbances and abnormalities of the T waves and RS T transitions

DIAGNOSIS In aortic stenosis a loud rough aortic systolic murmur an aortic systolic thrill absent or faint second aortic sound and a small delayed pulse suffice to justify the diagnosis The finding of calcification of the aortic valves on roentgenologic examination is confirmatory In the presence of mitral stenosis the diagnosis is often impossible to make

Those who have a calcific form of the disease have a good prognosis The outlook remains favorable so long as compensation is maintained Heart failure is usually fatal in this disease

Tricuspid Valvular Disease Organic tricuspid insufficiency is generally caused by rheumatic fever In some instances it may result from trauma or congenital malformation Occasionally a functional tricuspid stenosis originates from bacterial vegetations or thrombi Still more rarely tumors projecting into the tricuspid orifice cause an insufficiency The tumors may be primary myxoma or sarcoma or secondary metastatic lesions of hypernephroma sarcoma of the testis carcinoma of the thyroid or other neoplasms

The usual age of incidence is between 25 years and 40 years but occasionally the disease is present in persons more than 50 years of age Women are more commonly affected than men

Tricuspid insufficiency and stenosis are usually a part of rheumatic fever The right ventricle may be normal in size or only slightly hypertrophied but usually it is both dilated and hypertrophied The venae cavae are dilated Systemic venous distention occurs early The liver spleen and kidneys reveal chronic passive congestion Cardiac cirrhosis of the liver is frequent

The dynamics of tricuspid lesions may be modified and are often dominated by those of an associated mitral or aortic valvular disease

SYMPTOMS The symptoms of tricuspid valvular disease are those of right sided heart failure with systemic venous engorgement

EXAMINATION On examination there is cyanosis which is more pronounced in those who have tricuspid and mitral stenosis combined than in patients who have mitral stenosis alone Jaundice may be present

In tricuspid insufficiency the negative venous wave is replaced by the so-called positive venous pulse This positive venous pulse is often visible as a broad systolic wave ascending the neck along the course of the internal jugular vein up to the ear

In tricuspid insufficiency there is usually a palpable pulsation of the liver during ventricular systole The systolic hepatic pulse is elicited as an expansile sensation when one palm is held over the posterolateral surface and the other anteriorly over

the liver Practice is required to attain sufficient skill to elicit this pulse Hepatic enlargement is constant There often is ascites

In primary tricuspid stenosis and in tricuspid insufficiency peripheral edema may be absent In cases of functional tricuspid insufficiency edema may be absent but it is usually present

Percussion reveals an enlarged heart The right border may reach the right midclavicular line indicating a constant and definite cardiac enlargement

The findings revealed by examination in tricuspid valvular disease are often determined by the associated mitral stenosis or other valvular disease When tricuspid insufficiency is manifest there may be a reverberatory movement of the thoracic wall due to a pulsating liver and ventricular systoles

The tricuspid lesion may produce a systolic murmur over the lower end of the sternum or adjacent to it This murmur is indistinguishable from a mitral murmur When tricuspid stenosis is present a diastolic murmur may be heard which may be accompanied by a thrill However these findings are usually those of mitral stenosis or of mitral and aortic disease

The electrocardiogram reveals a right axis deviation and inversion of the T wave in lead III and often also of the T wave in lead II There may be wide notched and high voltage P waves when there is sinus rhythm Auricular fibrillation is common

Diagnosis The presence of systemic venous congestion cyanosis ascites and valvular heart disease is sufficient for a presumptive diagnosis of tricuspid heart disease The roentgenogram reveals clear lungs and enlargement of the right atrium

Functional tricuspid insufficiency often signifies the end stage of chronic valvular heart disease or pulmonary disease and therefore is always serious

Differentiation of tricuspid valvular heart disease from constrictive pericarditis is difficult In constrictive pericarditis the heart is not enlarged and evidence of organic cardiovalvular disease is absent

Organic tricuspid valvular disease may be compatible with life for a long period during which the only discomforts if any result from venous engorgement punctuated by recurring ascites

Pulmonic Valvular Disease Pulmonary insufficiency is classified as congenital and acquired A clinical separation of these two classes of pulmonary valve lesions frequently cannot be made in the adult

Functional or relative pulmonic insufficiency occurs as the result of a high pressure in the pulmonary artery The commonest cause of organic pulmonary insufficiency is pulmonary emphysema and rarely bacterial endocarditis involving the pulmonic valve Bacterial endocarditis of the pulmonic valve is usually not accompanied by other valvular lesions

In pulmonic insufficiency there is dilatation of the pulmonary artery hypertrophy of the right ventricle with regurgitation of blood from the pulmonary artery

The symptoms of pulmonic insufficiency are always dominated by those of the primary disease Dyspnea and cyanosis may be intense Polycythemia is observed in the cyanotic patients and hemoptysis is frequent

Examination reveals the findings of the primary disease There may be a Graham Steell murmur in the second or third left interspace in the presence of mitral stenosis and accentuation of the second pulmonic sound

Roentgenologic examination discloses evidence of right ventricular hypertrophy and often a dilatation of the pulmonary artery

The diagnosis is rarely made clinically

Cor Pulmonale Hypertrophy of the right ventricle of the heart or cor pulmonale occurs in acute subacute or chronic form The acute form is commonly caused by large pulmonary emboli The subacute form frequently is due to metastatic carcinomatous infiltration of the perivascular lymphatics and of the pulmonary arterioles occasionally the subacute lesion follows embolism of the smaller pulmonary vessels Similar changes in the right ventricle have been found in sickle cell anemia attended by multiple dissemi

nated thromboses of the smaller pulmonary arteries Chronic cor pulmonale is often associated with enlargement of the right side of the heart in association with chronic obstructive emphysema silicosis bronchiectasis tuberculosis bronchial asthma silico tuberculosis kyphoscoliosis and pulmonary arteriolar sclerosis In silicosis and bronchiectasis cor pulmonale occurs oftenest in association with peribronchial and perivascular fibrosis rather than with the nodular fibroses In tuberculosis the changes in the right ventricle usually follow extensive sclerotic lesions in the lungs Thoracic deformities kyphoscoliosis and secondary emphysema seem to be factors in the production of pulmonary hypertension and hypertrophy of the right ventricle Pressure by tumor on the main pulmonary artery organized vascular thrombi schistosomiasis with parasitic invasion of the walls of the pulmonary vessels and diminution in the caliber of the vascular channels lead to increased resistance to the flow of blood and hypertrophy of the right ventricle

A congenital or acquired cardiac lesion with arteriovenous fistulae may cause pulmonary congestion and hypertension

Cor Pulmonale Due to Pulmonary Emphysema (*Emphysema Heart*) Arteriosclerotic changes in the larger pulmonary arteries as well as in the small arteries and arterioles of patients who have advanced emphysema are more frequent and more severe than in normal persons of similar age

Advanced pulmonary emphysema with considerable arterial oxygen unsaturation requires a greater cardiac output In response to the anoxemic stimulus to the bone marrow there is a development of polycythemia which is an attempt to restore the normal hemoglobin content of oxygen despite the diminution in percentage of oxygen saturation

When virulent bronchopulmonary infection complicates pulmonary emphysema there is inadequate oxygenation of tissue and right ventricular strain and failure ensue Bronchial asthma causes cor pulmonale indirectly by first producing an obstructive emphysema Pulmonary tuberculosis of an advanced degree like obstructive emphysema may produce both right ventricular hypertrophy and congestive heart failure

Hypertrophy and dilatation of the right ventricle are present in those who have severe kyphoscoliosis The significant type of kyphoscoliosis is situated in the lower cervical and upper thoracic regions or in the midthoracic region with the curvature convex to the right Men are predominantly affected In most cases the cause of kyphoscoliosis is hereditary and congenital However tuberculosis of the spinal column rickets poliomyelitis and osteoarthritis are the causative agents in a great many cases

In kyphoscoliosis in either the congenital or acquired form the thoracic cavity is shortened the diaphragm elevated and the volume of the thoracic cavity and consequently of the lungs is diminished The lung on the side of the deformity is compressed atelectatic and eventually becomes fibrotic

Symptoms Rarely are the symptoms of sufficient distinctiveness to cause more than a suspicion of chronic cor pulmonale Pulmonary emphysema exists as such with or without complications A progression of the disease is complicated by bronchitis or asthma or both A pulmonary circulatory insufficiency may exist in a compensated stage from the time of inception of the emphysema

During the compensated stage of pulmonary heart disease from emphysema there are symptoms of pulmonary hypertension in addition to those of asthma The exertional dyspnea of emphysema is more marked and there may be dyspnea at rest Occasionally cyanosis is intense and associated with polycythemia In the decompensated stage of pulmonary heart disease dyspnea cyanosis fainting vertigo somnolence and occasionally precordial pain are present

Examination Pulmonary hypertension may be suspected from accentuation of the second pulmonic sound and right ventricular hypertrophy suspected from a prominent impulse in the epigastrium There may be roentgenologic evidence of dilatation of the pulmonary arteries and of the right ventricle

When there is a *decompensated stage of pulmonary heart disease*

and cyanosis are intensified Associated with intense cyanosis are polycythemia hepatic enlargement systemic venous congestion and peripheral edema Peripheral edema and occasionally ascites are present

The circulation time is normal in uncomplicated pulmonary emphysema even during an attack of bronchial asthma and it remains normal during the compensated stage of chronic cor pulmonale When intrapleural pressure is increased so is the venous pressure

The electrocardiogram of chronic cor pulmonale may not disclose any abnormality or it may reveal the changes due to a vertical position of the heart depression of the diaphragm and right ventricular strain

DIAGNOSIS The diagnosis of pulmonary heart disease is more definite when right sided heart failure is associated with primary pulmonary disease Hepatic enlargement and subcutaneous edema in addition to elevation in venous pressure are indicative of right ventricular failure

The history and the results of physical and roentgenologic examinations of the thorax will usually reveal the presence and nature of the pulmonary disease and exclude primary cardiovalvular disease The diagnosis of compensated cor pulmonale requires demonstration of right ventricular enlargement indicated by the roentgenologic examination

DISEASES OF THE HEART DUE TO DISTURBANCES OF INNERVATION OR PSYCHIC CONTROL

Neurocirculatory Asthenia or Cardiac Neurosis This condition is known also as irritable heart disordered action of the heart and effort syndrome

Neurocirculatory asthenia is a syndrome of psychogenic or neurogenic origin simulating organic heart disease from which it often cannot be differentiated immediately There are complaints of dyspnea precordial pain palpitation and exhaustion

These patients often give a history of nervous breakdowns indecision prolonged illnesses and an intolerance for the vicissitudes of daily life This disorder however may occur in any person if there is imposed a sufficient amount of exhaustion and emotional trauma

Neurocirculatory asthenia may occur at any age in persons of either sex but it seems to be commonest in young men The hereditary and environmental factors may be important A similarity of somatic type of those who have the disorder is often noticeable They are of an asthenic build with a long and narrow thorax

In some of those who have neurocirculatory asthenia a serious illness has occasioned a long withdrawal from physical exercise Incident to the withdrawal there was time in which anxieties in regard to ultimate recovery seized the patient With subsequent illnesses the anxiety and prolongation of disability tend to recur In some patients the anxiety will recur in the absence of other illnesses

SYMPTOMS These patients complain of difficulty in breathing precordial discomfort palpitation vertigo hyperhidrosis weakness gastrointestinal disturbances and often headache On questioning it will be found that they are fearful of some indescribable danger and impending illness In addition to these complaints it will soon be evident these patients are irritable restless and unable to sleep There are present anorexia slowed processes of thought and feelings of confusion

A characteristic shortening of breath described as an inability to take a deep breath occurs while at rest or during exercise The severity of the dyspnea is disproportionate to the degree of exercise and to the objective disturbances in ventilation

Fatigability a sense of profound fatigue during the early morning hours and in some great exhaustion follow physical exertion A cardiac disorder which confines a patient to rest in bed for 2 or more months without stirring around is usually neurocirculatory asthenia

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